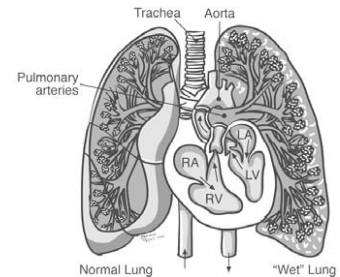


Heart Failure

The term congestive heart failure is often used to describe all forms of heart failure. Congestion (the build-up of fluid in the lungs or lower limbs) is just one feature of the condition and does not occur in everyone. Heart failure is a chronic condition characterized by the heart's diminished ability to pump blood effectively. Children with Barth syndrome are at an increased risk of developing heart failure. Small areas of damage may occur over years, damaged muscle tissue is replaced by scar tissue. The heart compensates for the failure in two ways:

- By increasing the number of beats
 - This helps pump more blood out of the heart and increases circulation.
- By enlarging and increasing the thickness of the heart muscle fibers
 - This helps the heart to increase its pumping strength by contracting with greater force.

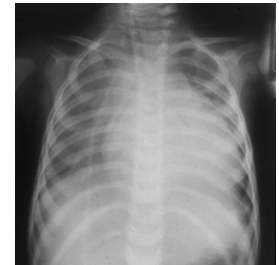


The heart can over-compensate for a period of time; however, eventually these compensatory mechanisms actually contribute to the progression of the disease by causing the walls of the heart to enlarge too much. The enlargement of the heart walls means less blood can flow through the chambers.

Signs and Symptoms of Heart Failure:

Babies

- Poor feeding. Requires longer feedings, more frequent feedings or tires easily with feedings
- Difficulty gaining weight and sudden weight gain
- May notice puffiness of the feet, face or hands
- Breathing fast, especially with feeding
- A big/distended abdomen (a big liver)
- Rapid heartbeat, sometimes visible to the family/naked eye. Sometimes parents report feeling child's heartbeat when they hold the baby.
- Excessive sweating with feeds
- More irritable
- Noticeable fatigue
- Inability to get his breath noticeable by muscle pulling or retracting between ribs and the breastbone
- Noisy breath sounds
- Fewer wet diapers, less urinating
- Chronic hacking or wet cough



Young children

- Similar to aforementioned symptoms
- Reduced exercise capability, having to stop and rest with exercise.
- Inability to keep up with peers; need to stop to catch their breath.
- Swelling of ankles (dependent edema) at bedtime, but unusual.

Teenagers

- Similar to aforementioned symptoms
- Palpitations because of the association with rhythm problems.
- Dizziness or fainting, especially during or after exercise.
- Sometimes, belly pain after eating, but this is very non-specific.

Heart Failure Fact Sheet

The Barth Syndrome Foundation www.barthsyndrome.org

6/1/2005 ~ Barry J. Byrne, MD, PhD; Arnold W. Strauss, MD; Carolyn T. Spencer, MD

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Glossary of Terms

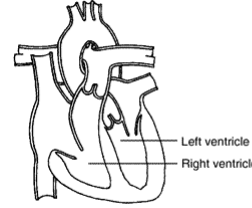
- **Angiotensin converting enzyme (ACE) inhibitor** is a drug used to decrease pressure inside blood vessels.
- **Arrhythmia** is an abnormal rhythm of the heart.
- **Atrium** is the chamber of the heart that collects blood returning from the rest of the body. In all vertebrates but fish, there are two atria, left and right. The right atrium collects deoxygenated blood from the body and passes it to the right ventricle. The left atrium collects oxygenated blood from the lungs and passes it to the left ventricle.
- **Beta Blockers** are drugs slow the heart rate and may be used to treat both heart failure and arrhythmia. They are considered anti-hypertensive drugs that limits the activity of epinephrine, a hormone that increases blood pressure.
- **Bradycardia** is an abnormally slow heartbeat.
- **Cardiac arrest** is a sudden stop of heart function. See also "sudden death."
- **Cardiac catheterization** is a procedure in which a thin, hollow tube is inserted into a blood vessel. The tube is then advanced through the vessel into the heart, enabling a physician to study the heart and its pumping activity.
- **Cardiomyopathy** is disease of the heart (cardio) muscle (myo) disease (pathy):
- **Dilated cardiomyopathy** is a disease of the heart muscle that leads to enlargement of the heart's chambers, robbing the heart of its pumping ability.
- **Hypertrophic cardiomyopathy** is a disease of the heart muscle causing thickening of the heart walls, interfering with the heart's ability to fill with and pump blood.
- **Coenzyme Q10 (Ubiquinone)** is a fat-soluble vitamin-like substance present in every cell of the body and serves as a coenzyme for several of the key enzymatic steps in the production of energy within the cell.
- **Congestion** is an abnormal fluid accumulation in the body, especially the lungs.
- **Defibrillator** is an electronic device designed to shock the heart back to regular rhythm.
 - **AED = Automatic External Defibrillators** are external devices that can be used by minimally trained individuals in emergency situations to deliver a shock to reset the heart that is fibrillating.
 - **ICD = Implantable Cardioverter Defibrillators** are battery operated devices that monitor and if necessary correct an abnormal heart rhythm by automatically sending electric charges to the heart.
- **Digitalis** is a drug used to increase the force of the heart's contraction and to regulate specific irregularities of heart rhythm. (Also known as Digoxin or Lanoxin)
- **Diastolic** refers to the relaxation phase of the heart's pumping cycle.
- **Diuretic** is a drug that helps eliminate excess body fluid; usually used in the treatment of high blood pressure and heart failure. (Also known as Furosemide or Lasix.)
- **Dyspnea** is a shortness of breath.
- **Echocardiography** is a test that bounces sound waves off the heart to produce pictures of its internal structures.
- **Edema** is an abnormal fluid accumulation in body tissues.
- **Electrocardiogram (EKG or ECG)** is a measurement of electrical activity during heartbeats.
- **Electrophysiology Study (EPS)** is a special type of cardiac catheterization used to evaluate arrhythmia. An **ablation** is a procedure used to eliminate abnormal electrical pathways in the heart that may produce arrhythmia. An ablation may occur in conjunction with an EPS if indicated.
- **Electrophysiologist (EP)** is a sub-specialist in cardiology that specifically investigates the rhythm and rhythm disturbances that causes the heart to pump blood. The EP may order specialized tests such as signal average ECG, Microvolt T-wave alternans, EPS and many other studies to investigate rhythm disturbances of the heart.
- **Endocardium** is the smooth membrane that covers the inside of the heart.
- **Holter monitor** is a special type of ECG monitor that evaluates the heart rhythm over 24 hours.
- **Idiopathic** means the cause is unknown.
- **Loop Monitor** is a battery operated heart monitor implanted just below the skin but should not to be confused with the ICD. The loop monitor is an event monitor that can be "downloaded" during the visit with the doctor to see if any abnormal rhythms occurred during a period of time. The loop monitor can last for as long as a year and in some cases longer.
- **Myocardium** is the heart muscle tissue.
- **Regurgitation** is the back-flow of blood through an insufficient valve.
- **Septum** is a muscle wall in the heart separating the chambers.
- **Sudden death event** is a cardiac arrest caused by an irregular heartbeat. The term "death" is somewhat misleading, because some patients survive.
- **Systolic** refers to the contraction phase of the heart's pumping cycle.
- **Tachycardia** is an abnormally fast heartbeat.
- **Ventricles** are the two lower chambers of the heart.
- **Ventricular fibrillation (v-fib)** is a rapid, irregular quivering of the heart's ventricles, with no effective heartbeat.

Heart with Hypertrophic Cardiomyopathy



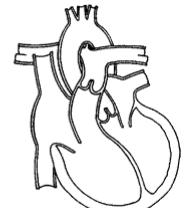
Growth and arrangement of muscle fibers are abnormal. Heart walls thicken, especially in the left ventricle.

Normal Heart



Heart chambers relax and fill, then contract and pump.

Heart with Dilated Cardiomyopathy



Muscle fibers have stretched. Heart chamber enlarges.

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