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INTRODUCTION

The term “cardiomyopathy” refers to a diseased state of the heart involving abnormalities of the muscle fibers, which contract with each heart beat. **(Colan et al., 2007).**

It can be primary when the muscle fibers themselves are abnormal as in genetic causes. It is also a secondary problem when the muscle fibers are normal but affected by other diseases that have secondary damaging effects. **(Colan et al., 2007).**

According to the pediatric cardiomyopathy registry, the majority of diagnosed patients are under 12 months followed by children 12 to 18 years old. **(Colan et al., 2007).**

There are 4 main types of cardiomyopathy & other unclassified types which have different clinical features & different lines of treatment. These types are: dilated, hypertrophic, restrictive & arrythmogenic right ventricular cardiomyopathy. **(Elliot p et al., 2008).**

This disease is associated with a significant risk for anesthesia , in addition, cardiac arrest under anesthesia has been attributed to undiagnosed cardiomyopathy. **(Elliot p et al., 2008).**

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Care of these patients is complicated by the fact that there are different forms of cardiomyopathy that have different anesthetic management goals. The main aim is maintaining the patients baseline hemodynamic variables of preload, heart rate, contractility & afterload. **(Kawaraguchi Yet al., 2002).**

With the emergence of new diagnostic tools, together with advances in cardiac imaging and improved treatment modalities such as ventricular assisted devices, the anesthetic management of these cases is evolving. **(Blauwet LA et al., 2010).**

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AIM OF THE WORK

The purpose of this essay was to present the different types of pediatric cardiomyopathies and their systemic effects that may interfere with anesthesia and provide guidelines for their anesthetic management.

CLASSIFICATION AND PATHOPHYSIOLOGY OF CARDIOMYOPATHIES

Cardiomyopathies in pediatrics are grouped into specific morphological and functional phenotypes. Theses phenotypes are :
Hypertrophic cardiomyopathy (HCM), Dilated cardiomyopathy (DCM), Restrictive cardiomyopathy (RCM), Arrythmogenic right ventricular cardiomyopathy (ARVC) & unclassified cardiomyopathies **(Elliot p et al., 2008).**

Each phenotype is then sub-classified into familial and non-familial forms. The familial subtype refers to the occurrence, in more than one family member, of either the same disorder or a phenotype that is (or could be) caused by the same genetic mutation and not to acquired cardiac or systemic diseases **(Elliot p et al., 2008).**

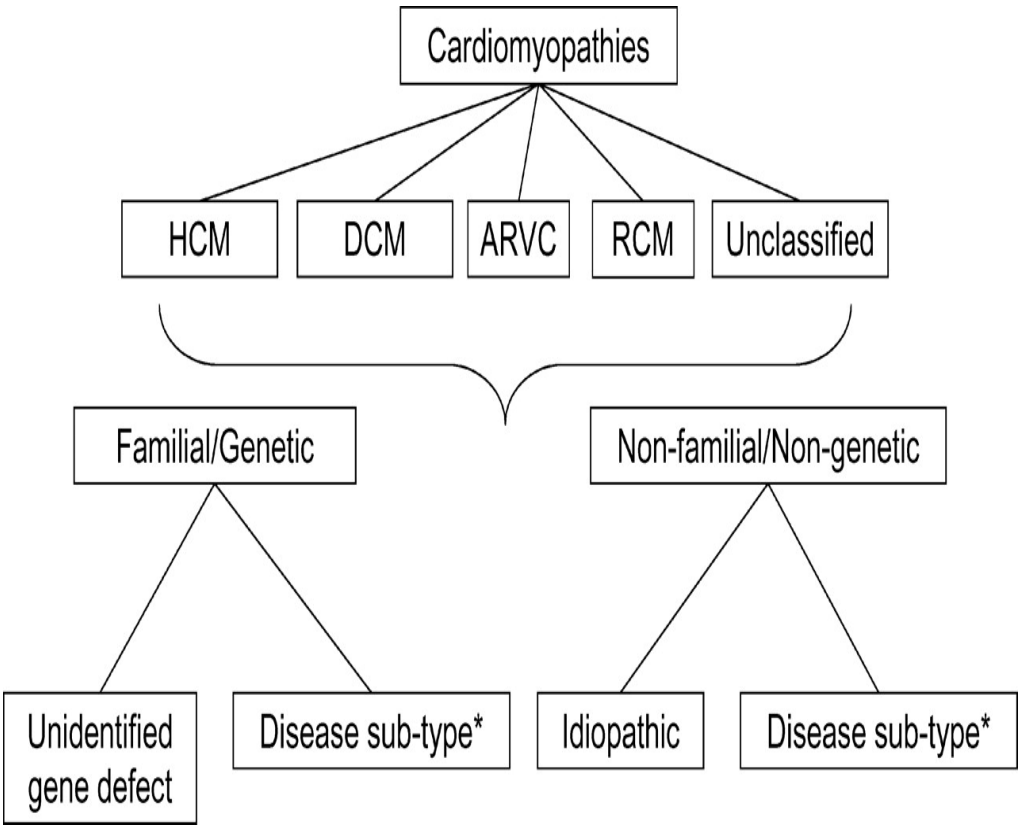
Most familial cardiomyopathies are monogenic disorders (i.e. the gene defect is sufficient by itself to cause the trait). A monogenic cardiomyopathy can be sporadic when the causative mutation is de novo, i.e. has occurred in an

individual for the first time within the family (or at the germinal level in one of the parents). In this classification system, patients with identified de novo mutations are assigned to the familial category as their disorder can be subsequently transmitted to their offspring. **(Elliot p et al., 2008)**

Non-familial cardiomyopathies are clinically defined by the presence of a cardiomyopathy in the index patient and the absence of disease in other family members (based on pedigree analysis and clinical evaluation). They are subdivided into idiopathic (no identifiable cause) and acquired cardiomyopathies in which ventricular dysfunction is a complication of the disorder rather than an intrinsic feature of the disease **(Elliot p et al., 2008).**

The expert panel of the American Heart Association has suggested that ion channelopathies and disorders of conduction should be considered as cardiomyopathies. This suggestion was predicated on the fact that these genetic disorders are responsible for altering biophysical properties and protein structure, thereby creating structurally abnormal ion channel interfaces and architecture **(Maron BJ et al., 2006).**

(Figure 1)



(McKenna WJ et al., 2004).

However, recent studies suggesting that genes encoding ion channels may be implicated in subgroups of patients with dilated cardiomyopathy (DCM), conduction disorders, and arrhythmias do not provide an argument for the redesignation of channelopathies as cardiomyopathies at the present time **(Alder E et al., 2005).**

Hypertrophic cardiomyopathy:

Simply defined by the presence of increased ventricular wall thickness or mass in the absence of loading conditions (hypertension, valve disease). It includes conditions with myocyte hypertrophy and those with increased left ventricular mass and wall thickness which are caused by interstitial infiltration or intracellular accumulation of metabolic substrates . **(Richardson p et al., 1996).**

This term can't be confined to the myocyte hypertrophy alone because it is frequently impossible to differentiate these two entities using non invasive techniques such as echocardiography or magnetic resonance imaging. In addition to, the patchy and complex nature of most myocardial pathologies means that this distinction can only be reliably made at post-mortem **(Richardson p et al., 1996).**

Causes

A-Familial **B- Non familial**

Familial causes include:

1- **Sarcomic protein metabolism** e.g. Cardiac troponin I, A-tropomyosin , Cardiac actin & ß myosin heavy chain.
2- **Glycogen storage diseases** e.g. Pompe disease & Danon disease.
3- **Lysosomal storage diseases** e.g. Anderson–Fabry disease & Hurler's disease.
4- **Syndromic HCM**: LEOPARD syndrome, Nonaan's syndrome , Swyer's syndrome & Friedreich's ataxia **(McKenna WJ et al., 2004).**

Non familial causes include:

Obesity, infants of diabetic mothers & amyloidosis **(McKenna WJ et al., 2004).**

Symptoms

Many patients with hypertrophic cardiomyopathy do not have symptoms. But if symptoms occur, at first they generally include:

- Shortness of breath (dyspnea).
- Chest pain or discomfort (angina).
- Fainting or near-fainting (syncope), especially with exertion.
- Heart palpitation, an uncomfortable awareness of the heart beating rapidly or irregularly **(Bernstein D et al., 2007).**

Sudden death may occur from the onset of ventricular tachycardia or other dangerous arrhythmia . A genetic factor appears to influence which patients with hypertrophic cardiomyopathy are more prone to sudden death **(Bernstein D et al., 2007)**

Other risk factors for sudden death include severe obstruction of the left ventricle, multiple fainting (syncope) episodes, recurring episodes of ventricular tachycardia, and an abnormal drop in blood pressure during exercise **(Bernstein D et al., 2007).**

Complications

- Atrial fibrillation is a common complication of hypertrophic cardiomyopathy so It can cause

blood clots to develop in the heart, which can break off and travel through the bloodstream (systemic embolism). This may cause a stroke or blocked blood flow to an arm or leg **(Bernstein D et al., 2007).**

- Heart failure is a common complication in which the heart's ventricles are not able to pump blood effectively enough to meet the body's needs for oxygen and nutrients. Common symptoms include fluid buildup (edema) in the legs, ankles, and feet; shortness of breath while lying down or exercising; and increased urination at night **(Bernstein D et al., 2007).**

Dilated cardiomyopathy:

It's defined by the presence of left ventricular dilatation and left ventricular systolic dysfunction in the absence of abnormal loading conditions (hypertension, valve disease) or coronary artery disease sufficient to cause global systolic impairment. Right ventricular dilation and dysfunction may be present but are not necessary for the diagnosis **(Elliot et al., 2008).**

Causes

A- Familial **B- Non familial**

Familial causes include:

1- **sarcomic protein metabolism** e.g. Cardiac troponin I, A-tropomyosin , Cardiac actin & ß myosin heavy chain.
2- **cytoskeletal genes** e.g. Dystrophin ,Epicardin & Desmin.
3- **nuclear membrane defects** e.g. Emerin. **(McKenna WJ et al., 2004).**

Non familial causes include:

- Myocarditis (infective/toxic/immune)
- Kawasaki disease
- Eosinophilic (Churg Strauss syndrome)
- Viral persistence
- Nutritional defects e.g. thiamine, carnitine, selenium , hypophosphataemia and hypocalcaemia
  (McKenna WJ et al., 2004).

Symptoms

Mild enlargement of the heart doesn't cause any

symptoms However, if the heart becomes very dilated, symptoms will develop. Symptoms may develop gradually or quite quickly, depending on how quickly the disease progresses . The symptoms are like those of heart failure such as, shortness of breath, retention of fluids (edema) and tiredness. Other common symptoms in infants are poor appetite and slow weight gain. Older children may show signs of poor exercise tolerance and gastrointestinal distress **(Jefferies JL et al., 2010).**

Complications

1- Dysrhythmias

There are various types of dysrhythmia that can be caused by dilated cardiomyopathy . Atrial fibrillation is the most common one that develops & may become permanent. Other arrhythmias may develop but not very common **(Jefferies JL et al., 2010).**

2- Blood clots

The blood flow through the heart becomes slower than normal, especially when the heart is very dilated. This can lead to small blood clots developing within the chambers of the heart. These may travel in the bloodstream and get stuck in arteries of the body. This may lead to a stroke if a clot gets stuck in an artery in the brain, or to other

problems. The risk of blood clots developing can be reduced greatly by taking an anticoagulant medicine such as warfarin **(Jefferies JL et al., 2010)**

3- Sudden death

Sudden collapse and death without any warning occurs rarely in patients with DCM. This is probably due to a severe arrhythmia which may develop suddenly. Medication and/or an implantable defibrillator may reduce this risk **(Jefferies JL et al., 2010).**

Restrictive cardiomyopathy:

Restrictive cardiomyopathies are defined as restrictive ventricular physiology in the presence of normal or reduced diastolic volumes (of one or both ventricles), normal or reduced systolic volumes, and normal ventricular wall thickness **(Kushwaha SS et al., 1997).**

Restrictive left ventricular physiology is characterized by a pattern of ventricular filling in which increased stiffness of the myocardium causes ventricular pressure to rise precipitously with only small increases in volume. Historically, systolic function was said to be preserved in RCM, but is rare for contractility to be truly normal.

Restrictive physiology can occur in patients with end-stage hypertrophic and DCM **(Kushwaha SS et al., 1997)**

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Causes

A-Familial **B- Non familial**

Familial causes include:

1- **Sarcomic protein metabolism** e.g. Cardiac troponin I, A-tropomyosin , Cardiac actin & ß myosin heavy chain.
2- **Familial Amyloidosis.**
3- **Glycogen storage diseases** e.g. Pompe disease & Danon disease.
4- **Haemochromatosis**.
5- **Anderson- Fabry disease**

(McKenna WJ et al., 2004).

Non familial causes include:

- Scleroderma
- Endomyocardial fibrosis
- Hypereosinophilic syndrome
- Idiopathic
- Drugs (serotonin, methysergide, ergotamine, mercurial agents, busulfan)

- Carcinoid heart disease
- Metastatic cancers
- Radiation **(McKenna WJ et al., 2004)**

Symptoms

Symptoms of heart failure are most common. Usually, these symptoms develop slowly over time. However, sometimes symptoms start very suddenly and are severe. Common symptoms are:

- Cough
- Difficulty in breathing
  - At night
  - Especially with exertion
  - When lying flat
- Fatigue, poor exercise tolerance
- Irregular or rapid pulse
- Loss of appetite
- Swelling of the abdomen
- Swelling of the feet and ankles **(Hare JM et al., 2007).**

Other symptoms may include:

- Chest pain
- Decreased alertness or concentration
- Failure to thrive
- Low urine production
- Shock (low blood pressure) **(Wexler RK et al., 2009).**