Pediatric Cardiomyopathy and Anesthesia

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Cardiomyopathy (CM) is defined by WHO as ‘a disease of the myocardium associated with cardiac dysfunction’ and is either: dilated, hypertrophic,restrictive, arrhythmogenic right ventricular, or unclassified

The incidence of paediatric CM is 4.8 per 100 000 infants and 1.3 per 100 000 children under 10 yr.

Of them:

Dilated CM 60%.
Hypertrophic 25%
Ventricular non-compaction 9%
Restrictive 2.5%
Arrhythmogenic right ventricular dysplasia 2%

The prognosis is poor.
40% of children presenting with symptomatic CM in the USA either receive a heart transplant or die within 2 y.

A significant part of CM remains undiagnosed by the surgery.

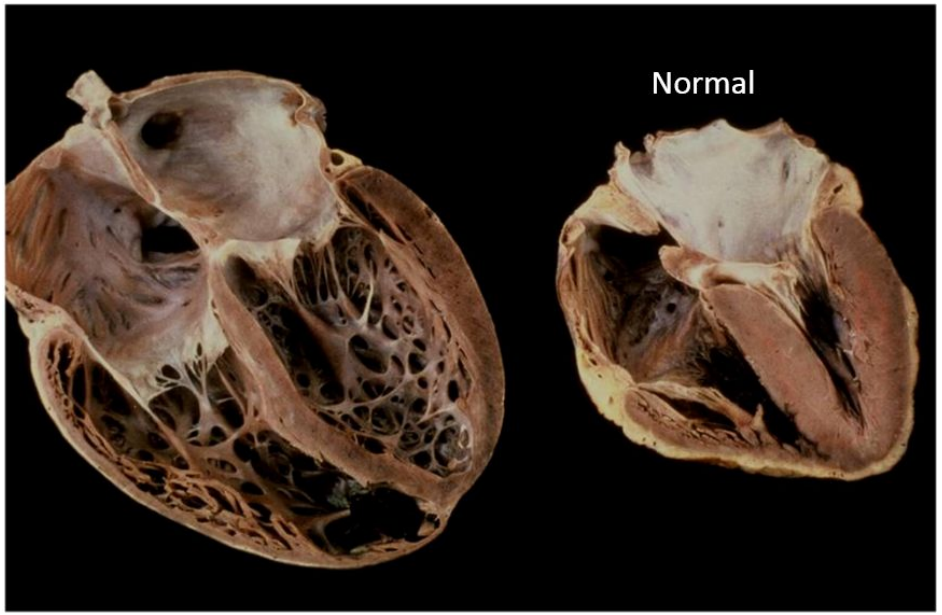
Children with either symptomatic or asymptomatic CM remain at significant risk of perioperative arrhythmia, cardiac arrest, and death

Dilated cardiomyopathy (DCM)

DCM, also called congestive CM, is characterized by dilatation and impaired contractility of one or both ventricles.

Annual incidence of DCM is 0.58 per 100 000 children

14% mortality rate in the 2 years after diagnosis

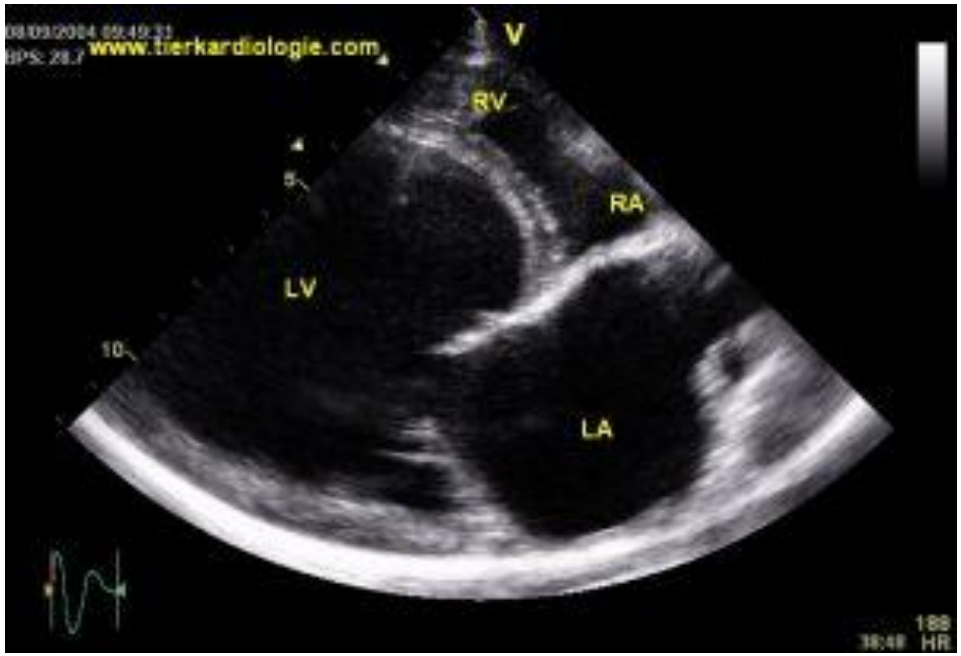


Ethyology:
congenital
Infection
Inflammation
metabolic or endocrine disease
malnutrition.
longstanding SVT
Idiopathic (66%)

7% of children who sustain burn injuries >70% BSA may develop a reversible DCM. This often presents 100 days after the injury. Inflammatory mediators?

Pathopfysiology of DCM

Biventricular dilatation
Systolic and diastolic myocardial dysfunction
Decreased EF
Decreased CO
Atrial filling pressure and LVEDP are elevated
Associated mitral and tricuspid valve regurgitation.
The dilated myocardium is potentially arrhythmogenic



Preanesthetic management of DCM

The enlarged heart extrinsic airway compression at the origin of the LMB.

Carefully adjusted CPAP overcomes the obstruction.

Most children have treatment with ACE inhibitors and β-blockers

Continue the treatment including the day of surgery, despite risk of hypotension.

Diuretics

Check volemia and potassium level

TEE is desirable to guide anesthesia

Anesthetic management of DCM

Optimization of coronary perfusion	Maintain adequate diastolic pressure Adequate preload
Maintenance of CO	Avoid cardiodepressive drugs (fentanyl/midazolam seems to be preferable) Low concentrations of Sevoflurane
Avoid increase of SVR	Avoid certain inotropes and ketamine as sole anesthetic
Inotropic support	Milrinon and dobutamine are preferable

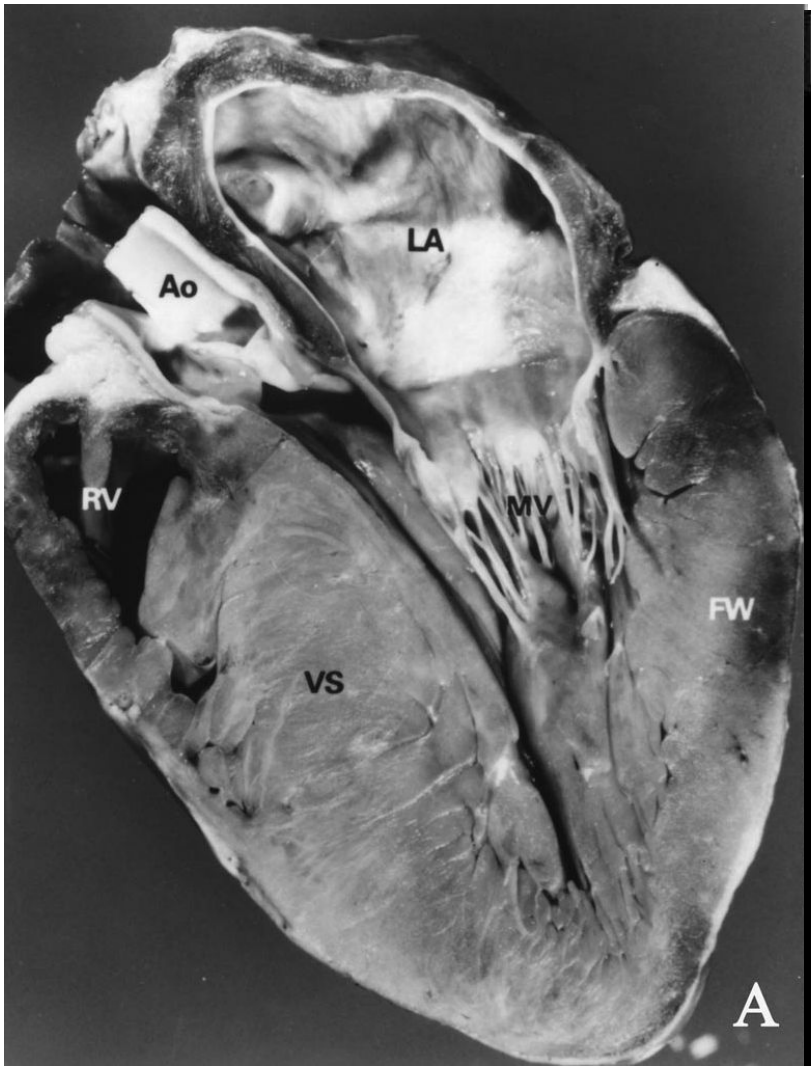
Hypertrophic cardiomyopathy HCM

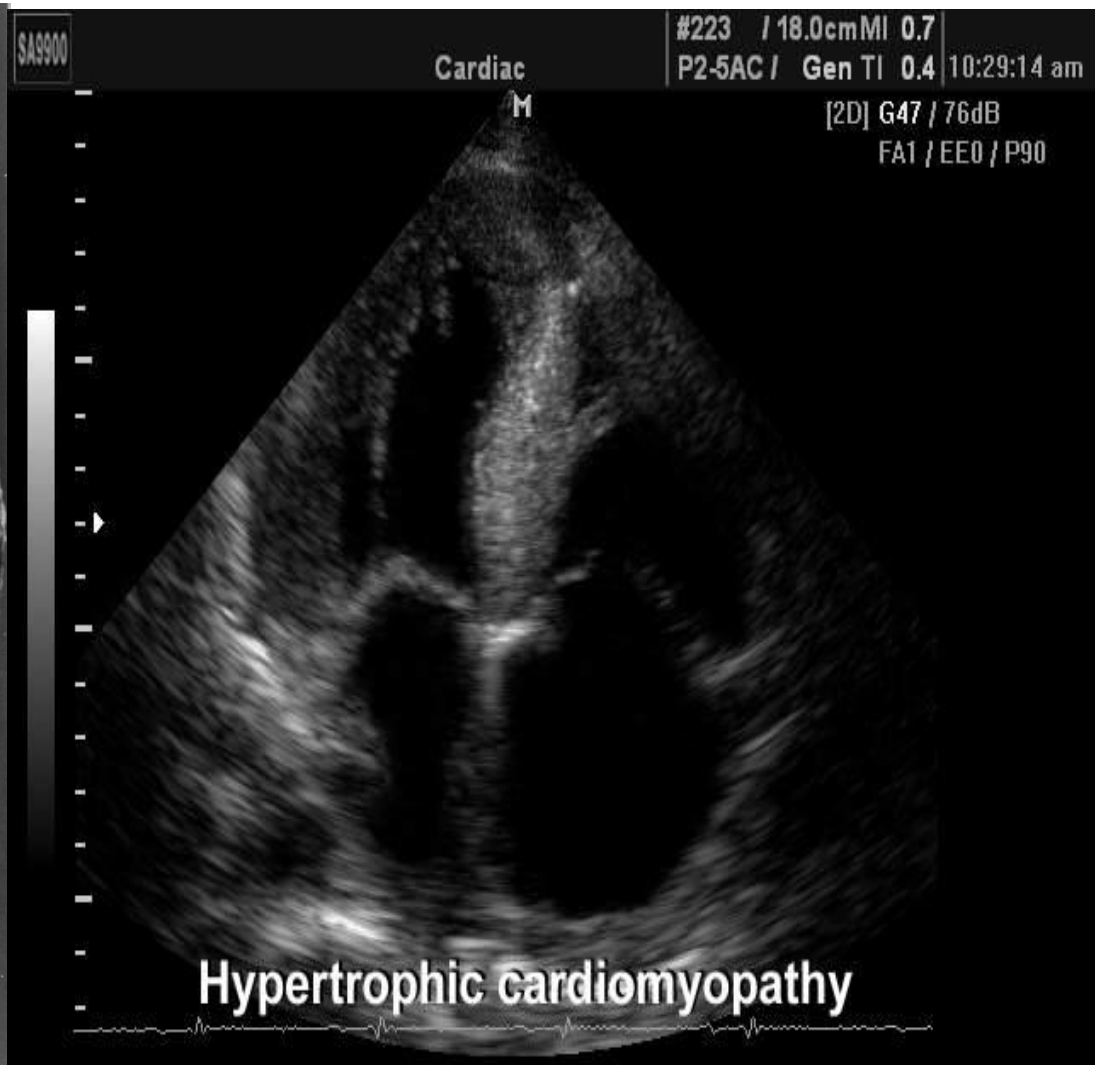
More common in adults, the incidence is low in children (5/1,000,000).
As patients can be asymptomatic, the diagnosis is often PM

A focal area of hypertrophy may also incorporate and surround a coronary vessel, so-called myocardial bridging □ significant coronary hypoperfusion □ risk of sudden death.

Pathophysiology of HCM

Asymmetric hypertrophy of septum & dynamic obstruction to LV outflow due to mitral valve systolic anterior motion and ventricular septal contact;





Hypertrophic_Cardiomyopathy_-_Echocardiogram_-_Sam (1).ogg

Factors affecting hemodynamics in patients with HCM

Improving	Deteriorating
Good preload/filling of LV, hypervolemia	Poor preload/ filling of LV, hypovolemia
Low contractility	High contractility
Bradycardia	Tachycardia, stress
High SVR	Low SVR
Low transaortic pressure gradient	High transaortic pressure gradient
Laying	Standing, Valsalva man.

Anesthetic management of HCM

Adequate preload	Aggressive fluid management prior and during surgery Aggressive correction of blood loss
Maintaining of elevated SVR	Phenylephrine
Avoid tachycadia, Decrease contractility	Continue β-blockers Perioperative β-blockers Anxiolytics Avoid ketamine, Ioflurane, propofol Avoid catecholamines Opioid based anesthesia + Sevoflurane Adequate pain control

Restrictive cardiomyopathy RCM

RCM - cardiac muscle disease resulting in
impaired ventricular filling with normal or decreased diastolic volume of either or both ventricles.
The condition usually results from increased stiffness of the myocardium

Progressive increase in PVR, due to blood flow to non-compliant LV, results in early mortality.

RCM has a 2 yr survival, once diagnosed, of 50%

ECHO diagnosis of RCM:
Small ventricles + massively dilated atria + elevated PAP

Severe changes in pulmonary vasculature prohibits heart transplant alone and a heart –lung transplant is the only alternative.

Restrictive cardiomyopathy
Anesthetic considerations

Due to stiffness of myocardium , CO depends on HR and preload.

Avoid bradycardia (fentanyl, penylephrine)

Maintain adequate preload

Fluid management, aggressive treatment of bleeding

Avoid increase of PVR

Avoid hypoxia, hypercarbia, hypothermia, elevated airway pressure

If inotropes needed

Milrinone and dobutamine

Arrhythmogenic right ventricular dysplasia/CM

Characterized by the gradual replacement of myocytes by adipose and fibrous tissue, it usually presents between the ages of 10–50 yr

ARVD/CM and long QT syndrome are the most common primary arrhythmic causes of SCD.

The inheritance of this disorder is autosomal-dominant

Pathologically, the free wall of the RV is replaced by fibro-fatty infiltration locuses for arrhythmias.

Arrhythmogenic right ventricular dysplasia/CM

Symptoms include palpitations, syncope, atypical chest pain, or dyspnea, but SCD may be the initial manifestation.

Of 50 autopsies performed for perioperative death, ARVD/CM was detected in 18 (36%). Four of the patients died on induction, 9 during surgery, and 5 within 2 h after surgery.

50% of patients have an abnormal ECG:
complete or incomplete RBBB,
QRS prolongation without RBBB,
epsilon wave immediately after the QRS in V1–V2,
T-wave inversion in V1–V3

Diagnosis:
1. ECHO: regional or global RV hypokinesis with or without dilatation
2. Angiography: RV wall anomalies in the absence of other structural heart defects
3. Histologically after an endomyocardial biopsy

Arrhythmogenic right ventricular dysplasia/CM Anesthetic considerations

Treatment with antiarrhythmics should be continued

Place external cardioversion/defibrillation pads on the chest before surgery

Avoid catecholamines

LA without adrenaline,
It is recommended to use lower doses of LA

Avoid tachycardia on induction

Adequate anesthesia, fentanyl.
Propofol is safe

Avoid reversal of NDMR block with atropine

Thank you!