WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 5.990
Volume 4, Issue 9, 520-528. Review Article ISSN 2277– 7105

REVIEW ON RESTRICTIVE CARDIOMYOPATHY

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Article Received on
22 June 2015,

Revised on 13 July 2015,
Accepted on 06 Aug 2015

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ABSTRACT

Restrictive cardiomyopathy is represented by RCM.RCM is a rare form of heart muscle disease that is characterized by restrictive filling of the ventricles. This occurs because the heart muscle is stiff and does not allow the ventricular chambers to fill with blood normally. This inability to relax and fill with blood results in a back up of blood in to the atria, lungs and body causing signs and symptoms of heart failure. RCM is least common in children accounting 2-5%of diagnosed RCM .RCM affects girls some what more than boys. In most of cases the cause is unknown (idiopathic) although a genetic cause is suspected in most of pediatric RCM. Diagnosis is made by exercise intolerance, ECG, cardiacbiopsy, cardiac catherization, increased pulmonary artery pressure.. Diuretics, beta blockers and occasionally after load reducing agents are most common medications in RCM.Anti coagulant medications are needed in children if there is a risk of blood clots inside the heart possibly leading to a stroke. No surgery has been effective in improving the RCM. Heart transplantation is only effective treatment in those with RCM.A high calories, electrolytes rich diet is preferred in RCM. Children with RCM are not allowed to play sports for long time due to risk of sudden collapse, heart failure. Prognostic studies show 20% RCM patients have risk of thrombosis.

KEY WORDS: RCM; Ventricular filling; Genetic cause; Heart transplantation.

ANATOMY

- Heart is a muscular organ about the size of a fist, located just behind and slightly left of the breast bone.
- The heart pumps blood through the network of arteries and veins called as cardiovascular system.
- The heart has four chambers, right atrium receives blood from the veins and pumps it to the right ventricle (fig: 1).
- Right ventricle receives blood from the right atrium and pumps to the lungs, where it is loaded with oxygen.
- Left atrium receives oxygenated blood from lungs and pumps to the left ventricle.
- The ventricle pumps oxygenated blood to the rest of the body .The left ventricle vigorous contractions create our blood pressure.

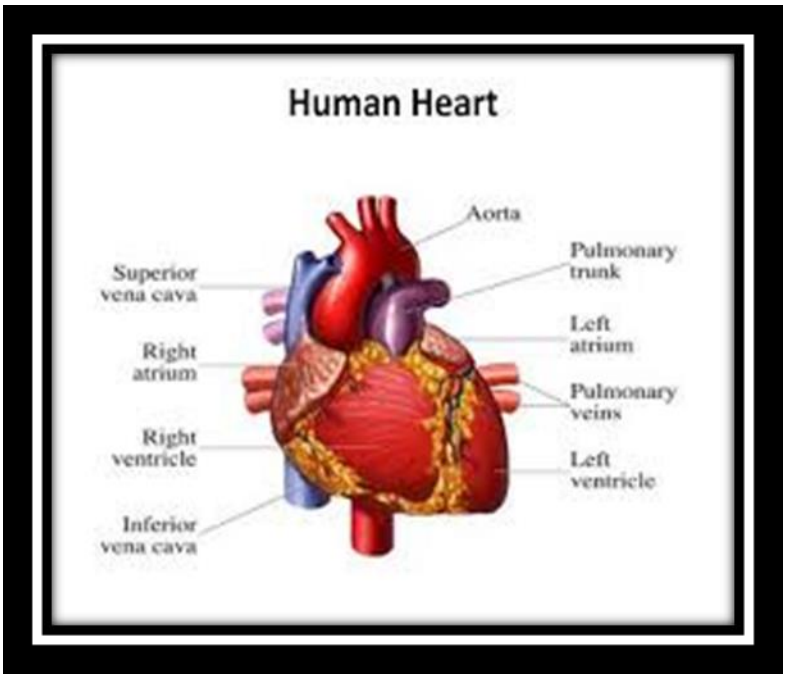

Fig: 1

INTRODUCTION

- In RCM the heart muscle has become stiff .This happens in lower heart chambers, which are called ventricles.
- Normally as your heart beats the ventricles expand fully and fill it with blood .This blood then gets pumped out to the body to supply it with oxygen and nutrients.

- But in case of RCM, the ventricles cannot stretch all the way to take in enough blood. So less blood is pumped out of the body. As it gets worse, heart failure develops (fig: 2).

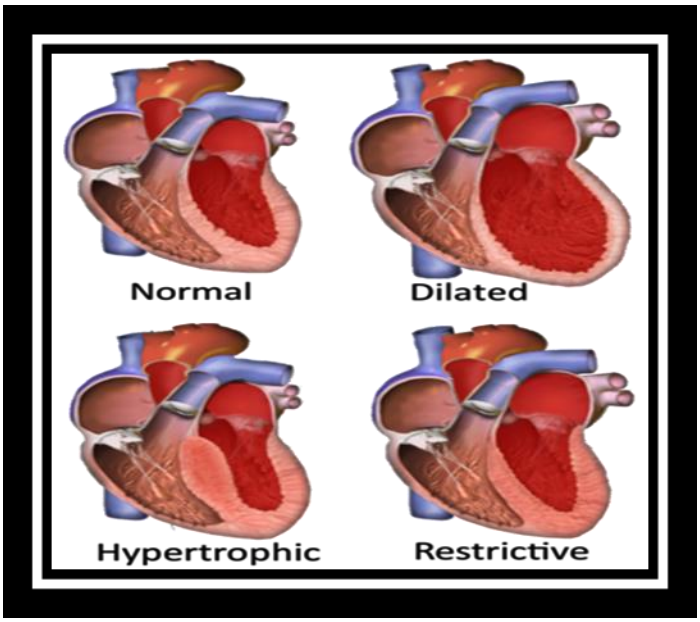

Fig: 2.

ETIOLOGY/CAUSES

- In most of the cases the cause is unknown
- Build up of proteins in heart muscle called as amyloidosis
- Build up of iron in the heart muscle called as hemochromatosis
- Condition of formation of masses (or) bumps in the heart and lungs (or)other organs called as sarcoidosis
- Radiation and chemotherapy treatments for cancer
- A type of cancer called carcinoid syndrome
- Build up of certain white blood cells that can lead to scarring .This can be caused by Loffler's syndrome (or) endomyocardial fibrosis
- Diseases that inherit from families like fabrys disease, gaucher disease can lead to RCM.

CLASSIFICATION

TYPE-A :(symmetric bi ventricular involvement)

- Large atria, normal /small ventricles
- Signs of pulmonary and venous congestion.

TYPE-B :(Asymmetric left ventricle involvement)

- Small left ventricle ,large left atrium, right heart chambers
- Signs of pulmonary congestion.

TYPE-C :(Asymmetric right ventricle involvement)

- Small right ventricle ,large right atrium
- Signs of venous congestion
- Decreased pulmonary and systemic pressures.

SIGNS AND SYMPTOMS

- Initial symptoms are absent (or) may have mild symptoms such as feeling of tiredness(or)weakness
- If heart gets weaker, it may leads to heart failure. When this happens ,you will feel other symptoms including,
- Shortness of breath ,especially with activity
- Tiredness
- Trouble breathing when you lie down
- Swelling of legs
- Chest pain
- Children with RCM may repeatedly experience asthma, respiratory tract infections
- Unfortunately, sudden death has been initial presentation in some patients.

DIAGNOSIS

- Diagnosis is made only after presentation with symptoms of RCM such as decreased exercise intolerance ,new heart sound (gallop),syncope(passing out) (or) chest pain with exercise.
- Biatrial enlargement and normal sized ventricles with normal (or) increased ventricular wall thickness are characteristic morphological features of RCM
- Microscopic examination reveals myocardial fibrosis, infiltration ,and myocardial scarring and myocyte heterotrophy (especially in idiopathic form).
- Diastolic dysfunction is a definable feature of RCM. Small sized rigid ventricles, high ventricular filling pressures.
- Elevated jugular pulse with prominent X and Y waves ,kussmaul sign and rising jugular pressure during inspiration, a palpable apical impulse, and murmurs of mitral /tricuspid

regurgitation, peripheral oedema, hepatomegaly, ascites and anasarca are diagnosed in advancing disease, new gallops.

- Restrictive pattern of ventricle filling often causes the symptoms like dyspnea,weakness,exercise intolerance and exertional chest pain.
- BNP levels are five times greater in restrictive cardiomyopathy, Normal BNP levels in the context of right sided heart failure should rise the suspicion of constrictive pericarditis.
- ECG displays non specific ST changes, low voltage, trial abnormalities,

and ventricular arrhythmias. Twelve-lead ECG recording from a patient with amyloidal infiltration of the heart. Note the low voltage in the precordial leads and a prolonged PR interval at 20 ms (shown in fig.3).

- ECG of children with RCM shows marked enlargement of atria, normal sized ventricles and normal heart function.
- Chest radiography shows an abnormal cardial silhouette caused by left atrial and bilatrial dilatation, evidence of pulmonary venous congestion (or)pericardial effusion.
- Cardiac catherization , cardiac biopsy are the other diagnostic procedures.
- CT, MRI scan discriminate between restrictive cardiomyopathy and constrictive pericarditis.

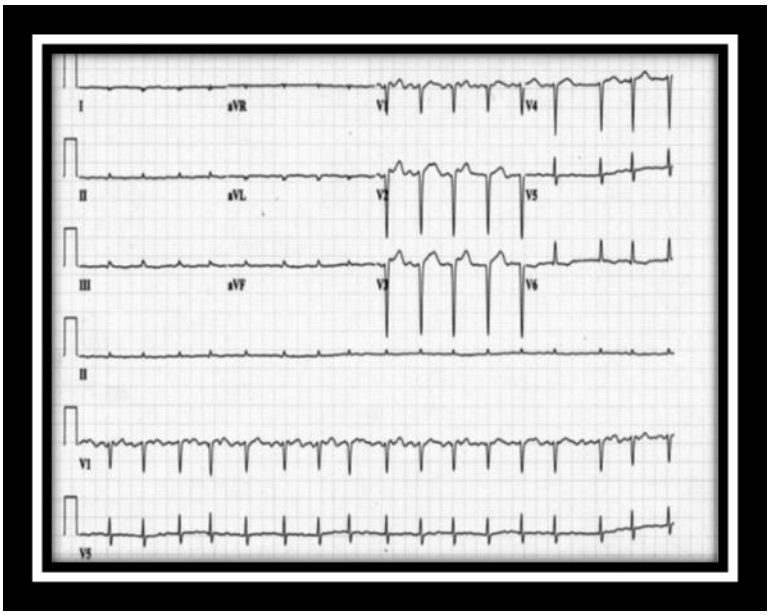

Fig: 3

MEDICATIONS

DIURETICS

- Commonly used are Furosemide, spironolactone, metolazone, bumetanide.

- These can be given orally (or) intravenously (IV).
- Dehydration ,excessive fluid loss is the major side effects.
- These reduce the fluid overload in lungs (or)other organs.

BETABLOCKERS

- Commonly used are carvedilol,metoprolol,propranolol,atenolol.
- Dizziness, low heart rate, low blood pressure ,fatigue, depression are major the side effects.
- These reduce the heart rate and increase the relaxation time of the heart.

ANTICOAGULANTS

- Commonly used are warfarin,heparin,and enoxaparin.
- Excessive brusing,bleeding from other minor skin injuries are the major side effects.
- These are mainly used in children with RCM because of risk of formation of blood clots leading to stroke.

HEART TRANSPLANTATION

- Since there are no proven effective therapies for children with RCM ,heart transplantation is the only intervention(fig:4).
- Especially preferred in case of fatal pulmonary hypertension.
- For children with RCM, heart transplantation can address both the abnormal heart function as well as associated pulmonary hypertension.
- Once the heart transplantation is done, other concerns arise, such as infection, organ rejection, coronary artery disease and side effects of medications.

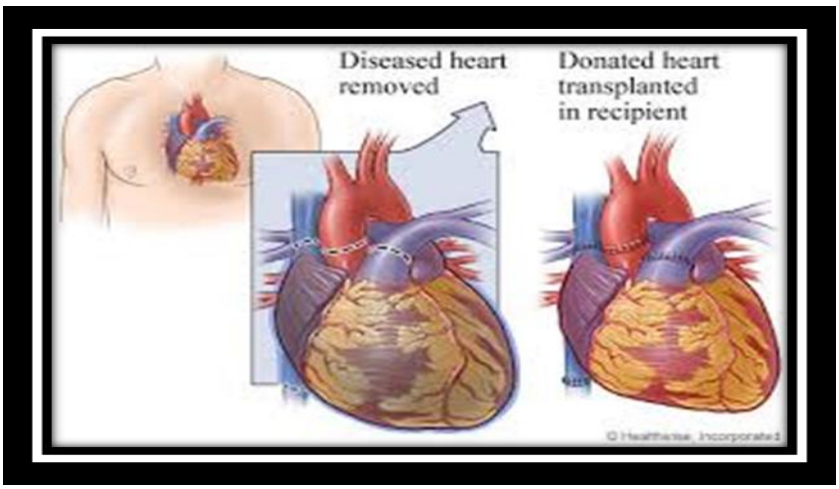

Fig: 4.

TO HELP YOUR SELF FEEL BETTER

- Limit how much salt (sodium) you get from food and drinks. When your heart cannot pump blood well, your body tends to hold on to extra salt and water. This leads to fluid buildup and swelling.
- Ask your doctor how much salt you can eat each day.
- Avoid processed foods. Limit potato chips, pretzels, salted nuts, processed meats and cheeses, pizza, canned soups, canned vegetables, olives, fast foods, and frozen dinners.
- Eat fresh or frozen fruits and vegetables instead of canned foods. Buy foods labeled“ low-sodium."
- Watch how much fluid you drink if your doctor tells you to. Ask your doctor how much is safe for you to drink.
- Weigh yourself each day to see if fluid is building up in your body.
- Find the right balance of rest and activity. You may need to limit exercise. Your doctor can help you find what is safe for you.
- Limit the amount of alcohol you drink.

WHAT DOES THE FUTURE HOLD FOR RCM?

- Slowly, progress is being made in our ability to diagnose RCM in both the clinical and molecular arenas. However, much additional research is needed in this field. Areas of research to be highlighted over the next decade include:
- Understanding of RCM as a disease process and the characteristics of the disease as they relate to outcome, which will lead to better management strategies;
- Increased clinical trials which will lead to new drug development and more effective therapies;
- molecular identification of novel genetic mutations as well as more precise diagnostic genetic testing/screening which will result in more accurate diagnosis.
- It is the expectation of the medical community that the data derived from exploring these avenues of scientific research will translate into a clinician’s ability to tailor medical therapy based on a given child’s precise diagnosis.
- Achieving this goal over the next couple of decades will represent a large milestone in the field of pediatric cardiomyopathy and will, hopefully, improve the ongoing care and prognosis of children afflicted with these heart muscle diseases.

CONCLUSION

It can be hard to treat restrictive cardiomyopathy because often the cause cannot be found. But if the cause is found and caught early, treatment can help. Medicines may be used to treat the cause. And treatment may limit some of the damage to the heart. But if the cause is hard to treat or not known, restrictive cardiomyopathy usually leads to heart failure over time.

REFERENCES

1. Ammash NM, Seward JB, Bailey KR, Edwards WD, Tajik AJ. Clinical profile and outcome of idiopathic restrictive cardiomyopathy. Circulation., 2000; 101: 2490–6.
2. Nihoyannopoulos P. Cardiomyopathies. In: Nihoyannopoulos P, Kisslo Jeds, Echocardiography. Springer; 2009.
3. Pepys MB. Amyloidosis. In: Weatherall DJ, Ledingham JGG, Warrell DAeds, Oxford Textbook of Medicine. Oxford: University Press; 1995; 2: 1512–24.
4. Benson MD, Uemichi T. Transthyretin amyloidosis. Amyloid Int J Exp Clin Invest., 1996; 3: 44–56.
5. Palka P, Lange A, Donnelly JE, Scalia G, Burstow DJ, Nihoyannopoulos P. Doppler tissue echocardiographic features of cardiac amyloidosis. J Am Soc Echocardiogr., 2002; 15: 1353–60.
6. Aroney C, Bett N, Radford D. Familial restrictive cardiomyopathy. Aust N Z J Med., 1988; 18: 877-8.
7. Fitzpatrick AP, Shapiro LM, Rickards AF, Poole-Wilson PA. Familial restrictive cardiomyopathy with atrioventricular block and skeletal myopathy. Br Heart J., 1990; 63: 114-8.
8. Branea I, Stanciu L, Tomescu M, Berinde L, Mancas S. The value of echocardiography in the early diagnosis of myocardial impairment due to connective tissue diseases. Med Interne., 1986; 24: 197-205.
9. Schurle DR, Evans RW, Cohlmia JB, Lin J. Restrictive cardiomyopathy in scleroderma. J Kans Med Soc., 1984; 85: 49-50.
10. Navarro-Lopez F, Llorian A, Ferrer-Roca O, Betriu A, Sanz G. Restrictive cardiomyopathy in pseudoxanthoma elasticum. Chest., 1980; 78: 113-5.
11. Bouchard A, Sanz N, Botvinick EH, et al. Noninvasive assessment of cardiomyopathy inMnormotensive diabetic patients between 20 and 50 years old. Am J Med., 1989; 87: 160-6.

12. Fein FS, Sonnenblick EH. Diabetic cardiomyopathy. Cardiovasc Drugs Ther., 1994; 8: 65-73.
13. Chew C, Ziady GM, Raphael MJ, Oakley CM. The functional defect in amyloidal heart disease: the "stiff heart" syndrome. Am J Cardiol., 1975; 36: 438-44.
14. Cueto-Garcia L, Tajik AJ, Kyle RA, et al. Serial echocardiographic observations in patients with primary systemic amyloidosis: an introduction to the concept of early (asymptomatic) amyloidal infiltration of the heart. Mayo Clin Proc., 1984; 59: 589-97.
15. Matsui Y, Iwai K, Tachibana T, et al. Clinic pathological study of fatal myocardial sarcoidosis. Ann N Y Acad Sci., 1976; 278: 455-69.
16. Perry A, Vuitch F. Causes of death in patients with sarcoidosis: a morphologic study of 38 autopsies with clinicopathologic correlations. Arch Pathol Lab Med., 1995; 119: 167-72.
17. Smith RL, Hutchins GM, Sack GH Jr, Ridolfi RL. Unusual cardiac, renal and pulmonary involvement in Gaucher's disease: interstitial glucocerebroside accumulation, pulmonary hypertension and fatal bone marrow embolization. Am J Med., 1978; 65: 352-60.
18. Renteria VG, Ferrans VJ, Roberts WC. The heart in the Hurler syndrome: gross, histologic and ultrastructural observations in five necropsy cases. Am J Cardiol., 1976; 38: 487-501.
19. Dervan JP, Ilercil A, Kane PB, Anagnostopoulos C. Fatty infiltration: another restrictive cardiomyopathic pattern. Cathet Cardiovasc Diagn., 1991; 22: 184-9.
20. Furth PA, Futterweit W, Gorlin R. Refractory biventricular heart failure in secondary hemochromatosis. Am J Med Sci., 1985; 290: 209-13.
21. Hillsley RE, Hernandez E, Steenbergen C, Bashore TM, Harrison JK. Inherited restrictive cardiomyopathy in a 74-year-old woman: a case of Fabry's disease. Am Heart J., 1995; 129: 199-202.
22. Olson LJ, Reeder GS, Noller KL, Edwards WD, Howell RR, Michels VV. Cardiac involvement in glycogen storage disease III: morphologic and biochemical characterization with endomyocardial biopsy. Am J Cardiol., 1984; 53: 980-1.