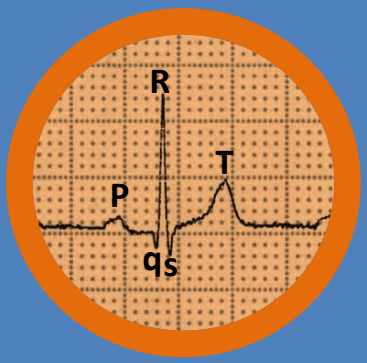


ECG Review : Cardiomyopathy

(For Academic Purpose only)

Dr. DURGA PRASAD KHAITAN

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The muscle thickens or stretches wide ,
Less strength to pump the blood aside …..

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The muscle thickens or stretches wide ,
Less strength to pump the blood aside …..

Knowledge and skill in the field of electrocardiography are constantly changing with the new researches and understanding.

With humble words I wish to say that some articles of my wirte-up on Cardiomyopathy are being covered in this book. It is only a step towards the vast ocean of knowledge. I may be excused for any error or omission.

With thanks and regards



DEDICATED
TO ALL THE
FELLOW COLLEAGUES

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DILATED CARDIOMYOPATHY (DCMP) : FROM PATHOPHYSIOLOGY TO ECG MANIFESTATION

ECG

DILATED CARDIOMYOPATHY (DCMP) : FROM PATHOPHYSIOLOGY TO ECG MANIFESTATION

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OUTLINE

Introduction

Dilated cardiomyopathy (DCMP) is considered as a symmetrical dilated left ventricle (LV) endowed with poor systolic contractile function ; right ventricle (RV) may also be involved as a companion.

Pathophysiological approach to DCMP

Basic Histopathology : myofibrillatory loss accompanied with interstitial fibrosis and T-cell infiltrates.

Compensatory mechanism in response to continuous decline in ejection fraction.

- ☐ Neurohormonal feedback, via activation of the sympathetic nervous system and the renin-angiotensin system.
- ☐ Frank-Starling law.

ECG manifestation (the reflection of histopathological changes)

- ECG traid – heart failure syndrome
- Assocaited fibrosis with conduction system defects
- ST segment depression and T-wave inversion
- Arrhythmias
- 'Lone' findings
- fragmented QRS (fQRS)
- Sudden cardiac death

Illustration by ECGs

- ECG No 1 (with typical findings)
- ECG No 2 (with fragmented QRS)

Take-Home Message

References

Dilated cardiomyopathy (DCMP) : From Pathophysiology to ECG manifestaion

A Narrative Review

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We have advanced from an era of descriptive cardioilogy to one of mechanistic conception. This sort of understanding proves to be better mirror to understand the nature of cardiac illness and a way better to targeted therapies.
In DCMP, the ventricle enlarges not out of strength, but out of necesssity – not beyond a point , increased volume no longer translated into effective output. The mechanistic of this pathophysiology can be explained mainly through two factors :

- **Neurohormonal feedback , via activation of the sympathetic nervous system and the renin-angiotension system , ultimately leads to increased ventricular wall stretch.**
- **Frank-Starling law offers grace only within a limits ; once overstretched , the myocardium loses its ability to respond what appears as a adaptation is often the beginning of decline.**

This is rightly said that the felicity in life depends upon how much we respond to 'Bio-physiological adaptation'.

1. Introduction

Dilated cardiomyopathy (DCMP) is considered as a symmetrical dilated left ventricle (LV) endowed with poor systolic contractile function ; right ventricle (RV) may also be involved as a companion. This pathophysiological entity suggests underlying heart failure syndrome with low ventricular ejection fraction.

This clinical pathophysiological entity is traditionally said to occur without coronary artery disease , hypertension , any significant valvular lesion or congenital heart disease. However , in contemporary practice there exists a distinct category which also includes ischemic injury as ischemic cardiomyopathy. The same also holds true with diabetic cardiomyopathy.

Approximately one-third of the cases are familial in origin , others are associated with chronic excessive alcohol intake or late autoimmune reaction to viral myocarditis. Some anti-neoplastic agents (doxorubicin , trastuzumab , imatinib) and multiple other risk etiologies like connective tissue disorders , heamochromatosis , sarcoidosis , muscular dystrophies , etc. should also be taken into consideration. Most cases of DCMP are idiopathic in nature.

In North-America and Europe , the incidence of symptomatic dilated cardiomyopathy is 20/100000 with a prevalence of 38/100000. Males are affected more compared to females almost with a ratio of 2:1. Such cases are also encountered not uncommonly in India. ECG is a commonly used diagnostic tool in cardiology which can also be applied to detect such cases with a specific set of findings with a moderate specificity but low sensitivity.

2. Pathophysiological approach to DCMP

This would be easier to grasp the electrocardiographic pattern in DCMP by exploring the associated pathophysiological process.

The basic histopathological changes show myofibrillatory loss accompanied with interstitial fibrosis and T-cell infiltrates. This is a slowly creeping pathological entity associated with left ventricular remodelling. There is a gradual increase in left ventricular end-diastolic and end-systolic volumes with consequent ventricular dilatation with its wall thinning resulting in an alteration in the contour of the heart , being more spherical - rather less elongated one. The continuous gradual decline in ejection fraction has been observed in DCMP.

Compensatory mechanism

With the progress of DCMP , two compensatory mechanisms are activated in response to continuous decline in ejection fraction.

- ☐ Neurohormonal feedback, via activation of the sympathetic nervous system and the renin-angiotensin system.
- ☐ Frank-Starling law

In the beginning this compensatory mechanism tends to compensate the decline in ejection fraction by increasing cardiac output and DCMP may remain asymptomatic but ultimately this is overwhelmed by the reduced response to this protective mechanism i.e. , it then becomes detrimental with intravascular volume expansion which usually leads to progressive dilatation of the ventricular system with the symptoms of heart failure.

Compensatory mechanism

- ☐ Neurohormonal feedback, via activation of the sympathetic nervous system and the renin-angiotensin system ⟶ An increase in systemic vascular resistance , with an increase in venous pressure ⟶ an increase in the volume of the blood to the ventricles ⟶ increased stretching of the ventricular wall.
- ☐ Frank-Starling law
  The stroke volume of the heart increases in response to this new equation by stretching the ventricular wall in response to an increased blood volumes to the ventricles ⟶ this initiates compensatory increase in cardiac output.

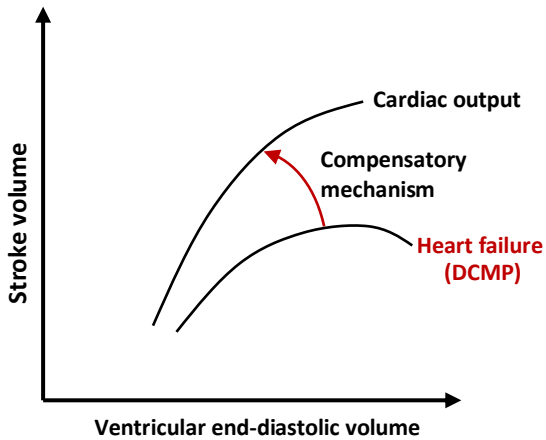


Fig. 1.1

☐ **Frank-Starling law**

3. ECG manifestation

ECG changes in DCMP might be said to be the reflection of its histopathological changes - a loss of functional muscle mass with fibrosis resulting in generalized low voltage in limb leads with poor R wave progression in chest leads. Due to the continuous operation of compensatory mechanism there might be relatively prominent QRS voltages in the chest leads.

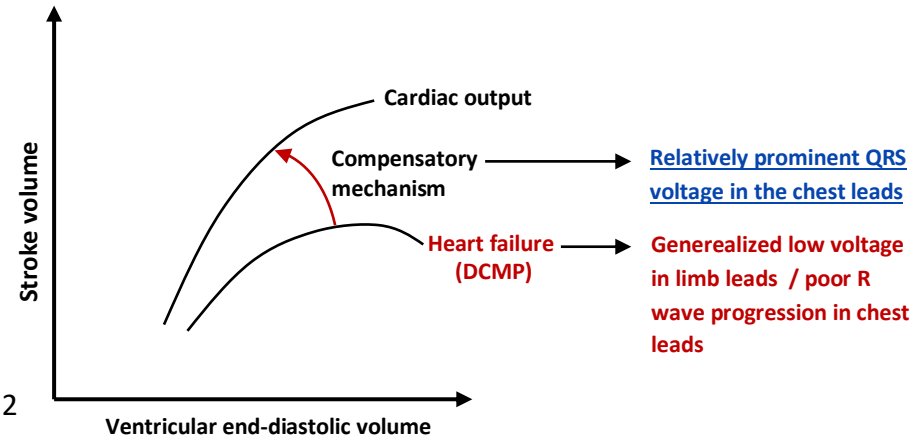


Fig. 1.2

This sketch illustrates how the underlying heart failure in DCMP with associated compensatory mechanism imparts its stamp on ECG changes. **This ECG triad, as mentioned below has moderate specificity but low sensitivity**. This pattern does not indicate whether heart failure syndrome is due to ischemic or non-ischemic insult.
The entire facts are put below to be reviewed **at a glance** :

ECG triad - heart failure syndrome (Goldberger's Triad Criteria)

1. Relatively low voltage in the limb leads – QRS in each of the six limb leads 8 mm or less in amplitude.
2. Poor R wave progression defined by QS / rS-type complex in lead V1-V4.
3. Relatively prominent QRS voltage in the chest leads usually measured as SV1 + RV5 or RV6 >35 mm.

These changes reflect increased LV dimension and posterior shifting of the cardiac vector in congestive heart failure. It is particularly noted in patient with severe LV dysunction (ejection fraction $\leq$ 20%).

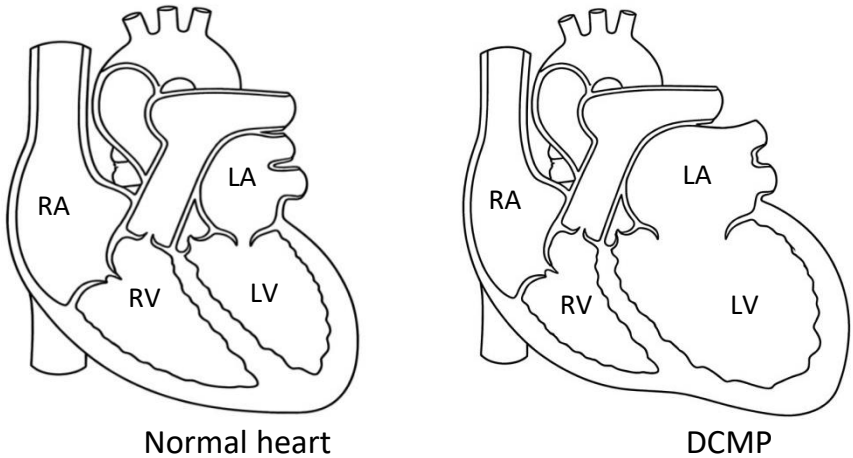


Fig. 1.3

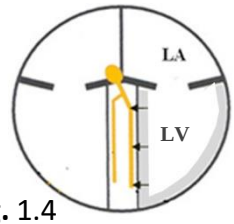
Associated fibrosis with the conduction system defects

AV nodal and interventricular conduction disturbances are usually present in most patients with advanced DCMP due to the associated fibrosis. They are manifested as :

- First degree AV block
- Left anterior fascicular block
- (Complete heart block is rare)
- LBBB pattern usually indicates diffuse myocardial disease with fibrosis

  A patient showing cardiomegaly of unknown cause with the presence of LBBB points towards the diagnosis of primary myocardial disease (the presence of RBBB is less common).

  The development of a new LBBB during follow-up is a strong independent prognostic predictor of all cause mortality.



Creeping interstitial fibrosis involving the left side of interventricular septum with impinging over the left bundle branch resulting in LBBB.

Fig. 1.4

- Diffuse non-specific QRS widening (>120 ms) – higher the QRS duration in patients with DCMP , higher is the morbidity and mortality.
- Abnormal Q wave mimicking anterior or inferior MI may be observed as a result of myocardial fibrosis (inert myocardial zone).

ST segment depression and T-wave inversion in left precordial and inferiorly oriented leads may point towards primary myocardial involvement , hence , indicate an important diagnostic criteria in a patient with clinically diagnosed DCMP.

Arrhythmias

A high incidence of supraventricular and ventricular arrhythmia has been reported in most of the cases in DCMP. Ventricular arrhythmias have been observed terminally. Supraventricular tachycardia occurs occasionally. Atrial fibrillation tends to develop late and it is a sign of structural disease progression (it can lead to sudden decompensation of the heart failure).

It has been documented that QRS duration $\geq$ 120 ms is a significant predictor of ventricular arrhythmias.

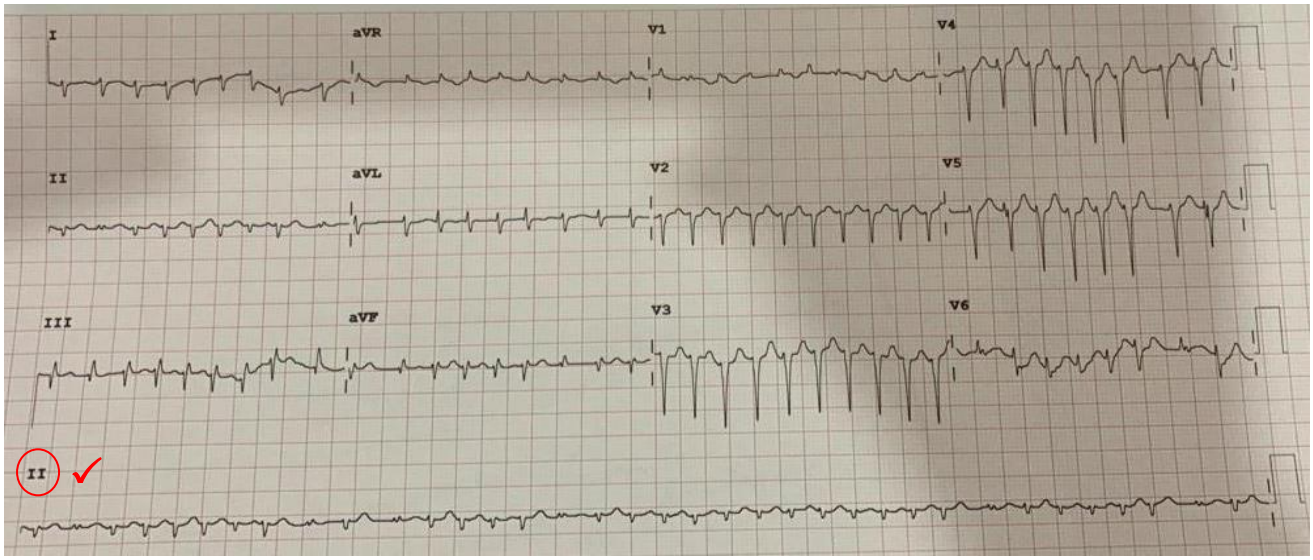
'Lone' findings

A few cases may show left atrial enlargement pattern alone , diffuse T-wave abnormalites or only sinus tachycardia without other ECG abnormalites.

At times **fragmented QRS** as a result of fibrosis may be observed on ECG.

Sudden cardiac death in DCMP can be caused by electromechanical dissociation (pulseless electrical activity) or ventricular arrhytmias. In electromechanical dissociation the person is unconsious having no palpable pulse and BP despite the presence of electrical waves on ECG. The patient has cardiac electrical activity but insufficient mechanical cardiac force to pump blood effectively.

4. Illustration by ECGs : **ECG No. 1** (Middle aged lady presenting with Breathlessness)



Source : Prof. Dr. A.N. Rai , Former Prof. & Head Medicine and Prinicipal ANMMCH , Gaya Bihar ; Chairman AIMS, Gaya



Chest X-ray PA view of the same patient :

- Spherical heart with cardiomegaly
- Lifting of the cardiac apex from the base is suggestive of the involvement of right ventricle as well.
- Upper pulmonary zone is having prominent pulmonary veins compared to the basal zone – suggestive of pulmonary vascular redistribution.

ECG findings :

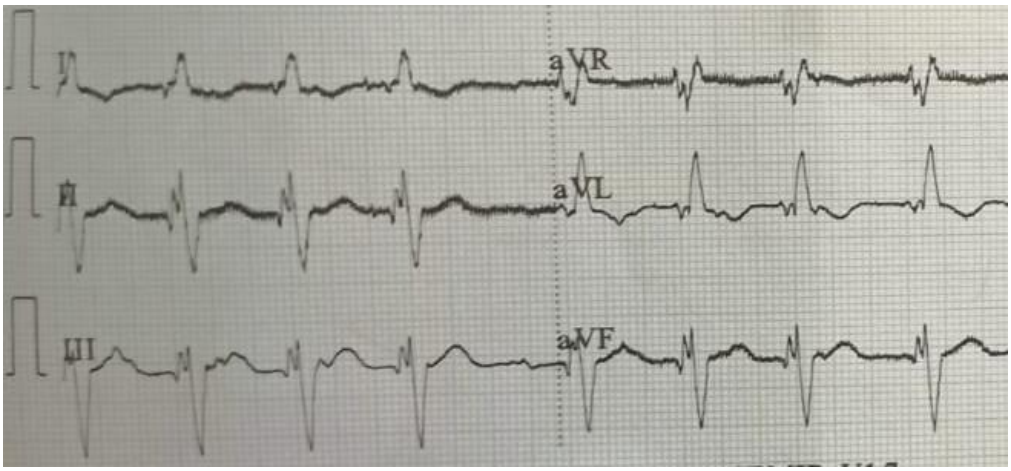
- Low voltage QRS in limb leads
- Poor R-wave progression in chest leads.
- Atrial fibrillation with fast ventricular rate (250 bpm) (please see the rhythm lead II : rhythm is irregularly irregular)
- North-west axis + 170^0 (negative polarity in limb leads II and aVF with +ve polarity in aVR)
- Occasionally fluctuating electrical alternans (please see lead V6)

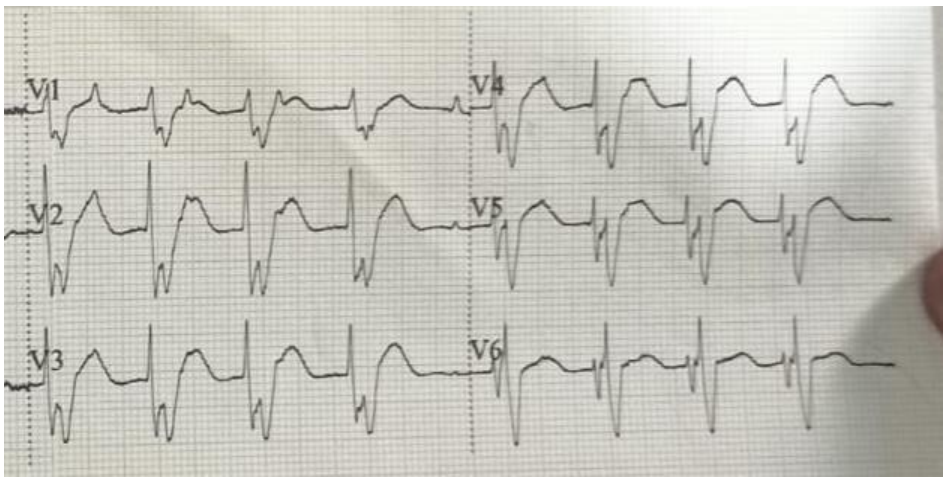
Discussion :

- The presence of low voltage in limb leads with poor R progression in chest leads are suggestive of DCMP.
- The presence of atrial fibrillation indicates this case to be long standing since this finding tends to develop late. It is also a sign of structural disease progression.
- Chamber enlargement alone is not sufficient to shift the axis to North-west direction. There must be anatomical distortion of His bundle and its branches to shift the axis dramatically towards this zone.
- Occasionally fluctuating electrical alternans is indicative of left ventricular dysfunction.

Final impression: DCMP with atrial fibrillation (mild pericardial effusion also demonstrated by cardiac echo in addition)

ECG No. 2





Source : CME INDIA on 25th May , 2022 by Dr. Sushil Kr. Goenka , Medical officer inchare , PHC , Birpur , Begusarai (Bihar)



Gross cardiomegaly with haziness in both lung fields

History :

62 years male , with breathlessness on effort (non-diabetic). LVEF was around 35%.

ECG findings :

- fragmented QRS : diffusely seen in both limb and chest leads having two notches in each R and S wave both, the distance in between notches being $\geq$ 40 ms.
- Sinus tachycardia (112 bpm) with first degree AV block (P is seen superimposed upon the top of T , most obvious in precordial leads).
- Left anterior fascicular block (Frontal QRS axis = -50^0)
- Presence of q over inferior leads II, III and aVF
- Poor R wave progression (R wave in V3 is not greater than R in V2 and R wave in V4 is not greater than R in V3)

It is to be noted that there is a lack of universal definition for **a preserved precordial R-wave progression** but common criteria include :

- R-wave > 2-4 mm in V3 or V4
- R-wave in V4>V3 or V3>V2
- R-wave in V3 > 3mm

Final impression : Most likely DCMP

5. Take-Home Message

- ☐ Dilated cardiomyopathy (DCMP) is considered as a symmetrical dilated left ventricle (LV) endowed with poor systolic contractile function ; right ventricle may also be involved as a companion. This clinical pathophysiological entity is said to occur in the absence of coronary artery disease , hypertension , significant valvular lesion or congenital heart disease. Most cases of DCMP are idiopathic in nature.

- ☐ For preliminary diagnostic purposes it should be kept in mind that the pattern of distribution of interstitial fibrosis is variable in nature and so the findings are also variable on ECG depending upon the loss of functional muscle mass with fibrosis.

- ☐ Earlier detection of the cases (by ECG modality) can slow the pathophysiological process and prevent heart failure syndrome , hopefully with increased life expectancy with a properly directed therapy.

    - Early DCMP (subtle stage) : Electrical signals are disturbed, but structural abnormality has not yet declared itself fully.
      Such as sinus tachycardia (may be mild), occasional PVCs, non-specific ST-T changes and possible early left atrial enlargement.
    - Advanced DCMP (established disease) : Now the ECG reflects not just dysfunction, but distorted architecture and unstable conduction also.
      Such as LBBB / wide QRS (very important marker) , Frequent ventricular arrhythmias (PVCs, NSVT, VT) , Clear atrial enlargement (often biatrial), Left axis deviation , more pronounced ST–T abnormalities , ECG triad – a heart failure syndrome (relatively low voltage in the limb leads , poor R-wave progression and relatively prominent QRS voltage in the chest lead) , etc.

      NB : High-yield clinical markers, such as a wide QRS complex (especially with LBBB), are suggestive of advanced remodeling associated with mechanical dyssynchrony, which results in further worsening of heart failure. Correction of this dyssynchrony by cardiac resynchronization therapy is an established treatment in this subgroup of patients.
      At times the pattern of fragmented QRS might be seen on ECG as an indicator of interstitial fibrosis.

- ☐ Worsening of the left ventricular function or increased incidence of ventricular arrhythmias burden can also be caused by associated development of new co-pathologies. Therefore, the associated presence of heart diseases like coronary artery disease , hypertensive heart disease , structural valvular disease or even acute myocarditis should be periodically ruled out during the follow-up. Even atrial fibrillation may deteriorate the functioning of the heart with further aggravation of the existing cardiac failure.

- ☐ Periodic ECG monitoring in patients with dilated cardiomyopathy (DCMP) may provide some insight into long-term prognosis, although its predictive value remains limited

☐ The echocardiography and cardiac MRI would be more useful in exploring the details in DCMP.

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LVH IN DCMP – A PUZZLE ON ECG

ECG

LVH IN DCMP – A PUZZLE ON ECG

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OUTLINE

Introduction

In dilated cardiomyopathy (DCMP), the left ventricle (LV) enlarges and its contractile function declines. Despite this dilatation, the myocardium often exhibits hypertrophy on ECG. This form of hypertrophy differs from that observed with pressure overload conditions such as hypertension or aortic stenosis.

Difference in LVH between DCMP and pressure overload conditions

- In DCMP , the heart exhibits eccentric hypertrophy
- The pressure overload conditions such as hypertension or aortic stenosis exhibit concentric hypertrophy

Pathology of LVH in DCMP

- ☐ Ventricular overload leading to LV dilatation , initially compensated by ventricular stretching as per **Frank-Starling law**
- ☐ Neurohormonal Activation (RAAS & Sympathetic Overdrive)
  - Myocardial hypertrophy as an adaptive mechanism
  - Fibrosis , which contributes to electrical remodelling

Concerned ECG changes with its basic reasoning

Illustration by ECGs

- ☐ **ECG Case 1 DCMP**
- ☐ **ECG Case 2 Degenerative Aortic Valvular Disease plus Diabetic cardiomyopathy (DCMP)**

Take Home Message

References

LVH in DCMP – A Puzzle on ECG

A Narrative Review

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Everyone loves solving puzzles from an early age. Even scientists find joy in unraveling complex puzzles , using their intellect to search out its solutions. It looks to be a very fascinating challenge that captivates the mind and sparks curiosity in everyone.

Dilated cardiomyopathy (DCMP) presents a diagnostic paradox ; the dilated heart exhibits hypertrophy. This discrepancy posses a compelling puzzle in the minds of those clinicians who are very much interested in cardiology and electrocardiography.

- ☐ **In dilated cardiomyopathy (DCMP), the left ventricle (LV) enlarges and its contractile function declines. Despite this dilatation, the myocardium often exhibits hypertrophy**
- ☐ **This form of hypertrophy differs from that observed with pressure overload conditions such as hypertension or aortic stenosis.**

Understanding the concept of left ventricular hypertrophy (LVH) in dilated cardiomyopathy is of paramount importance , as its management differs from that of classical LVH associated with pressure overload conditions.

1. Introduction (Keypoints)

- The basic histopathological changes in DCMP show myofibrillatory loss , accompanied with interstitial fibrosis and cellular infiltrates. Here the left ventricle (LV) is subjected to chronic overload due to systolic dysfunction (weakened contractibility) and increased end-diastolic volume (impaired ventricular ejection).

- Contrary to this dilatation , the myocardium often exhibits eccentric hypertrophy , different from concentric hypertrophy as seen with pressure overload conditions such as hypertension or aortic stenosis

- This eccentric hypertrophy in DCMP presents initially as compensatory phase and this is followed by the decompensatory phase as the heart failure ensues.

- There is a gradually declining left ventricular function due to global myocardial dysfunctioning (ejection fraction <40%).
  Patients usually present with symptoms of biventricular failure , e.g. fatigue , dyspnoea , orthopnoea , ankle oedema , etc.

- The documentation of precise incidence and prevalence rates of LVH in DCMP is lacking in the available literature. This lack of specific data may be due to the complex interaction between myocardial dilatation and hypertrophy in DCMP, further research is necessary for the purpose.

- Causes :

Genetic

Upto 25% are familial (primarily autosomal dominant , some types are X-linked)

Ischaemic

Dilated cardiomyopathy may occur here following massive anterior STEMI due to extensive myocardial necrosis and loss of contractility

Other causes :

- Late autoimmune reaction to viral myocarditis (coxsackie B)
- Chronic excessive alcohol intake including even cocaine.
- Some anti-neoplastic agents (doxorubicin , trastuzumab , imatinib)
- Autoimmune diseases like sarcoidosis and SLE
- Metabolic disorders such as diabetes mellitus and thyroid dysfunction
- Pregnancy (peripartum cardiomyopathy)
- Nutritional deficiencies : lack of essential nutrients particularly vitamin B1
- Idiopathic cases : **majority** cases occur without identifiable cause

NB : Long standing cases of Type 2 Diabetes mellitus (DM2) can lead to dilated cardiomyopathy : contributory factors might be chronic hyperglycaemia and increased fatty acid oxidation , microangiopathy , autonomic neuropathy impairing the myocardial contractility , oxidative stress with chronic low-grade inflammation.

- Patients with DCMP and LVH may respond differently to heart failure therapies. Regression of LVH with optimal therapy (ACE inhibitors, beta-blockers, and MRAs) correlates with better prognosis.
  Persistent LVH despite medical therapy may indicate poor response to reverse remodeling, necessitating CRT or ICD therapy in high-risk cases.

2. Difference in LVH between DCMP and pressure overload conditions

In DCMP , the heart exhibits eccentric hypertrophy –

The hypertrophied mass is laid down around the dilated ventricular cavity without its obliteration→ the overall myocardial mass is increased.

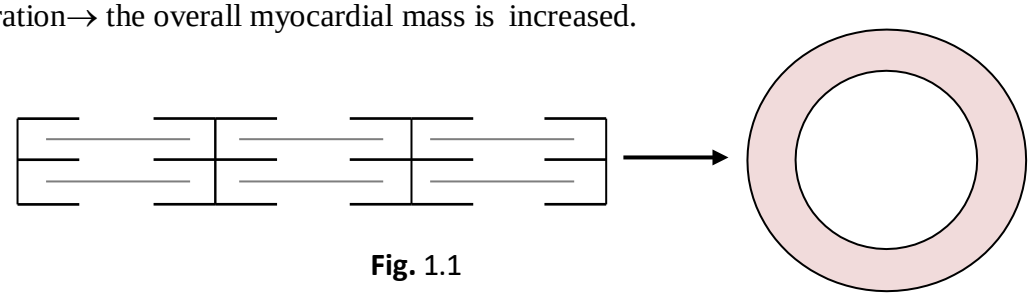


Fig. 1.1

Sarcomeres – the fundamental contractile units of muscle fibres , are added in series , i.e. , with end-to-end arrangement along the length of existing ones leading to enlongated muscle fibres with overall increase in ventricular mass.

This is known as eccentric hypertrophy.

The pressure overload conditions such as hypertension or aortic stenosis exhibit concentric hypertrophy.

Here the hypertrophied mass extending towards the centre of LV cavity , causing its significant obliteration.

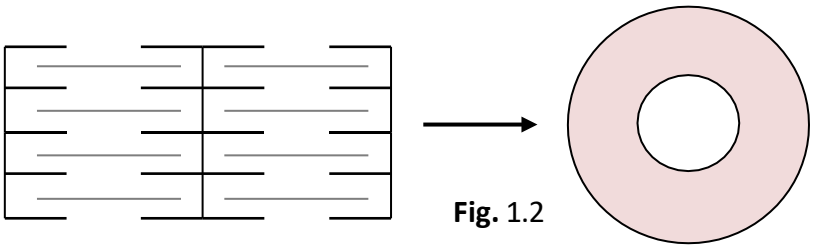


Fig. 1.2

Sarcomeres are added here in parallel , meaning thereby, new sarcomers are superposed upone each other leading to increased myocardial thickness with increased ventricular mass → thus obliterating LV cavity without its significant dilatation.

This is known as concentric hypertrophy.

Functional differences

DCMP	Pressure overload
Primarily associated with systolic dysfunction , with or without accompanying diastolic dysfunction.	Initially preserved systolic function , with diastolic dysfunction emerging over time.

3. Pathophysiology of LVH in DCMP

- ☐ Ventricular overload leading to LV dilatation , initially compensated by ventricular stretching as per **Frank-Starling law** (perceived as pseudo LVH on ECG)
- ☐ Neurohormonal Activation (RAAS & Sympathetic Overdrive)
  - Myocardial hypertrophy as an adaptive mechanism
  - Fibrosis , which contributes to electrical remodelling
- ☐ Progressive eccentric LVH with terminal decline in left ventricular functioning (Heart failure)
- ☐ **Secondary causes** : If any prior pressure overload , such as aortic stenosis or hypertension is superadded to the preexisting DCMP , then hypertrophy may supervene over the existing LV dilatation.

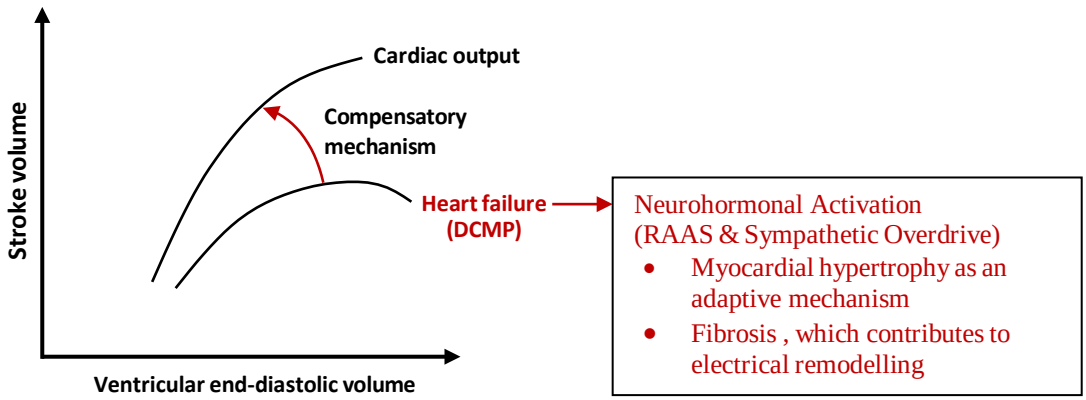


Fig. 1.3

Frank-Starling law

4. Concerned ECG changes with its basic reasoning

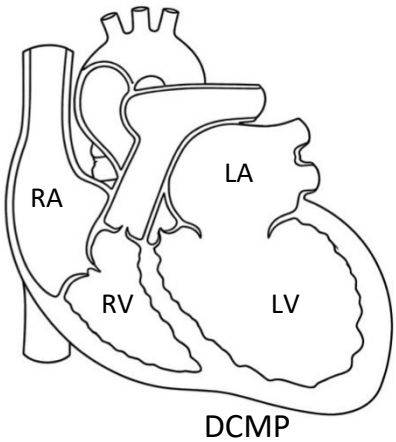


Fig. 1.4

Basic reasoning :

- LVH with LV dilatation (eccentric hypertrophy) with associated Fibrosis → shifting of electrical vector laterally and posteriorly :
  - Poor R-wave progression (PRWP)
  - Late QRS transition
  - Clockwise rotation
- In the presence of chamber dilatation , LVH pattern may be less pronounced than in pressure overload conditions.
- Interstitial fibrosis reflected in limb leads as relatively low voltage (limb leads are being away from the heart compared to those of chest leads).

Goldberger's triad is an electrocardiographic (ECG) pattern identified by Dr. Ary Louis Goldberger in 1982, associated with dilated cardiomyopathy (DCMP).

ECG triad

1. Relatively low voltage in limb leads (QRS voltage < 0.8 mV in each of the limb leads)
2. Poor R-wave progression across precordial leads: suboptimal increase in R-wave amplitude from leads V1 to V6 (e.g. R-wave ≤3 mm in lead V3)
3. Relatively prominent QRS voltage in the chest leads (LVH)

The quantum of these changes would depend upon the balance in between heart failure and compensatory mechanism operating therein.

This triad is considered highly specific for DCMP but is relatively insensitive, meaning its presence strongly suggests DCMP, yet its absence doesn't rule out this condition.
However , due to chamber dilatation , LVH pattern may be less pronounced than in pressure overload conditions.
Persistent LVH in DCMP suggests ongoing maladaptive remodelling.

LVH with associated fibrosis → slowed conduction across the myocardium

- Poor R-wave progression (PRWP) : This would be worthwhile to mention here that **as the LV enlarges , the heart's electrical vector shifts more laterally and posteriorly** , resulting in poor anterior forces leading to PRWP despite no infarction.
- Late QRS transition (normally V3-V4 but in DCMP V5-V6)
- Clockwise rotation due to LV dilatation and septal fibrosis , the depolarization wave moves more posteriorly , causing an apparent clockwise rotation

Conduction defects

- LBBB : This pattern usually indicates diffuse myocardial disease with fibrosis. The contributing factors to its causation are enumerated as below :

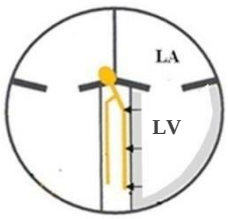


Fig. 1.5

- The mechanical stretch affecting the HIS-Purkinje system particularly the left bundle branch with slowing of the conduction therein.
- Creeping fibrosis disrupts conduction pathway especially in the left bundle branch , further promoting LBBB in DCMP patients.
- Delayed septal activation due to assocaited fibrosis may lead to ventricular dyssynchrony , further adding element to LBBB

The development of new LBBB during follow-up is a strong independent prognostic predictor of all cause mortality

- Intra Ventricular Conduction Delay (IVCD) without LBBB
- Other conduction defects like left anterior fascicular block , at times even first degree AV block may occur.

Arrhythmias

A high incidence of supraventricular and ventricular arrhythmias has been reported in most of the cases in DCMP. Ventricular arrhythmias have been observed terminally. Supraventricular tachycardia occurs occasionally**. Atrial fibrillation tends to develop late and it is a sign of structural disease progression** (it can lead to sudden decompensation with heart failure).

It has been documented that QRS duration $\geq$ 120 ms is a significant predictor of ventricular arrhythmias.

Peguero-Lo Presti and Sokolow-Lyon indices are commonly in use for diagnosing LVH

Due to shifting of electrical vector laterally and posteriorly with eccentric hypertrophy , Peguero-Lo Presti criteria seems to be more logical in diagnosing eccentric LVH

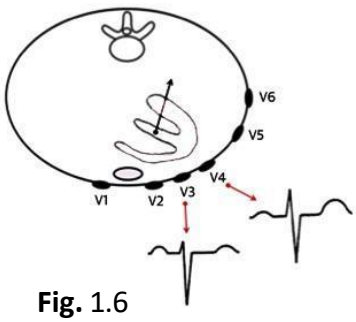


Fig. 1.6

Supplementary assessment, including imaging modalities like echocardiography or cardiac MRI, is highly recommended for precise evaluation.

Peguero-Lo Presti Criteria

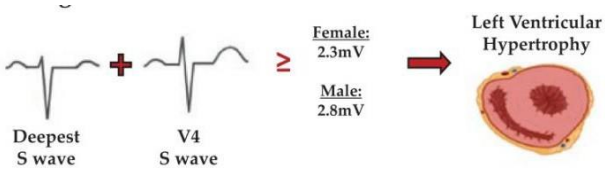


Fig. 1.7

Sensitivity: 47% to 55%, improving detection rates.
Specificity: Approx 72%, a higher rate of false positive , compared to Sokolow-Lyon.

Sokolow-Lyon criteria

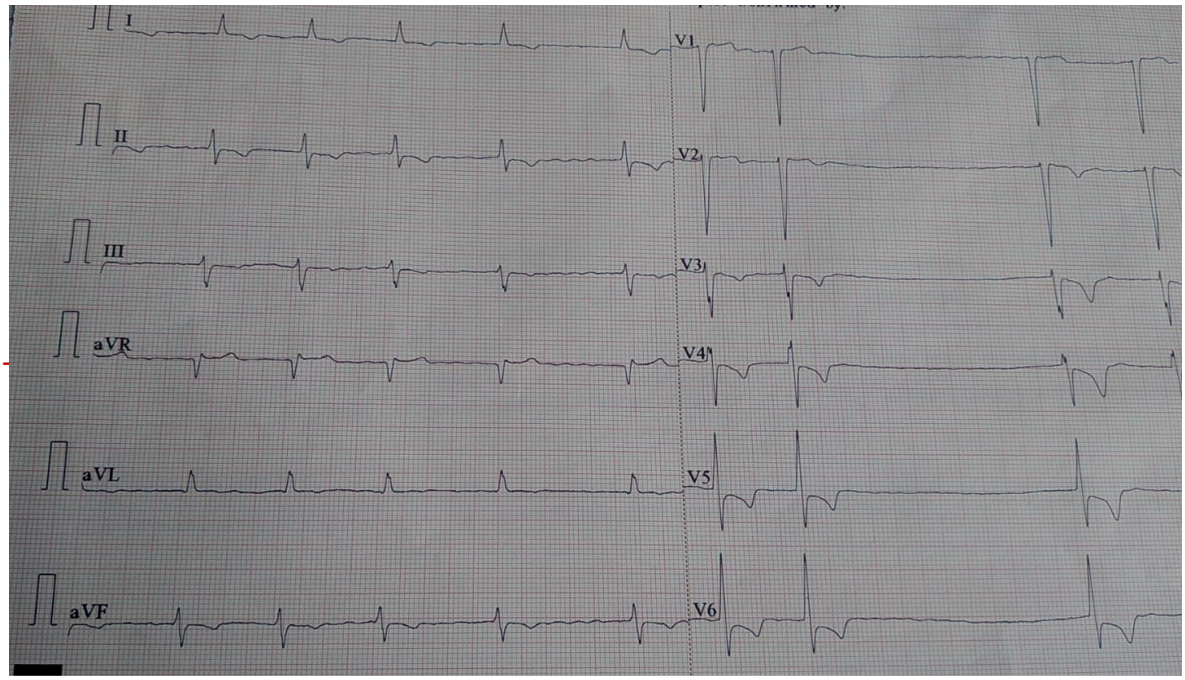
- S-wave depth in V1 + tallest R wave height in V5 or V6 > 35 mm
- R_{aVL} > 11 mm (with left axis deviation R in aVL should be >13 mm)

Sokolow-Lyon's index is the most used index, despite having the lowest sensitivity (20%) of all indexes. The specificity is high (>85%).

5. Illustration by ECGs

ECG Case 1 DCMP

50 Years old male with h/o Dysnoea



Source : Global Heart Rhythm Forum on 05.03.2025 **by Dr. R.K. Gupta** ,
Senior Consultant Physician , Yamunanagar , Haryana

ECG Findings	Comments
1. Atrial fibrillation (fine) with occasional long pause , lasting for 2.24 sec (see chest leads).	Atrial fibrillation tends to develop late and is a sign of structural disease progression. Longer pauses (over 4 seconds) or pauses with significant symptoms simulating that of SA block warrant medical attention.
2. Relatively low voltage in limb leads (QRS voltage < 8 mm in each of the limb leads)	Interstitial fibrosis reflected in limb leads as relatively low voltage
3. LVH by Peguero-Lo Presti Criteria (deepest S-wave in V2 19 mm + S wave in V4 10.5 mm = 29.5 mm with accompanying global non-specific ST/T changes)	LVH with associated fibrosis → slowed conduction across the myocardium. As the LV enlarges , the heart's electrical vector shifts more laterally and posteriorly , resulting in poor anterior forces leading to such changes
4. Triad • Poor R-wave progression (PRWP) : (R-wave<3 mm in lead V3 • Late QRS transition at V5 • Clockwise rotation	
5. fQRS (QRS fragmentation) at V3-V4 (obvious on zooming the tracings)	QRS fragmentation indicates fibrosis ± conduction defects

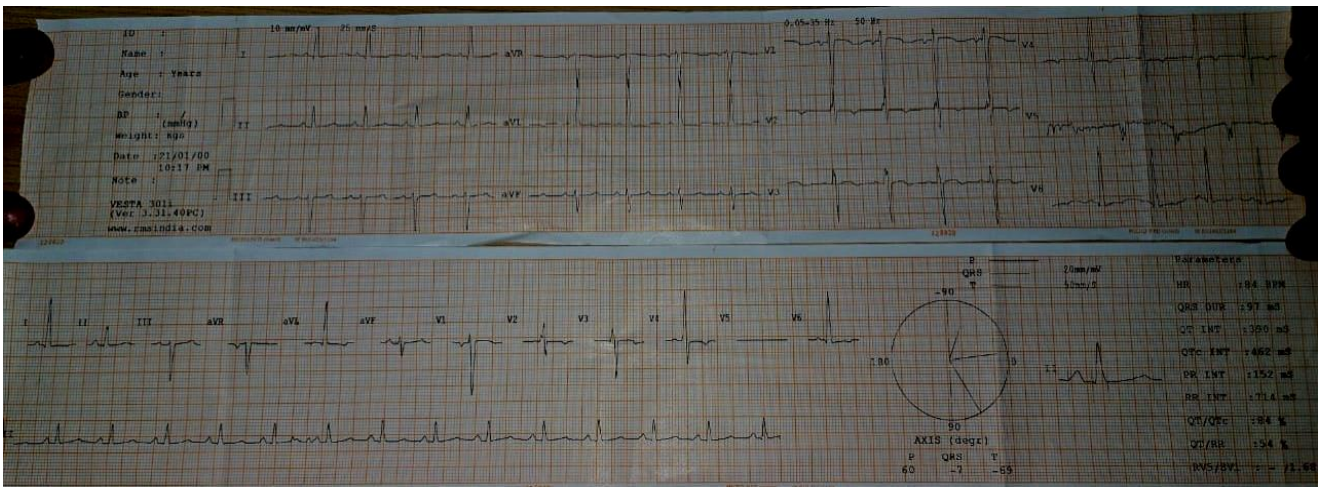
☐ **Cardiac Echo** : Severe left ventricular dysfunction with EF 25%

☐ **CAG** Normal

FINAL IMPRESSION : DCMP

ECG Case 2 Diabetic cardiomyopathy (DCMP) with degenerative aortic valvular disease

46 Years old female presented with SOB and swelling of the body , Normotensive , No past history of diabetes but random blood sugar 250 mg % with HbA1c 8.2



Source : CME INDIA on 05.03.2025 by **Dr. N.K. Singh** , Director , Diabetes and Heart Research Centre, Dhanbad , Editor , www.cmeindia.in

Findings on ECG :

- R in aVL > 11 mm with secondary ST/T changes in lead I and aVL , associated with somewhat axis tilting towards the left (frontal QRS axis -20 degree , with reciprocal mild ST elevation in lead V1-V2)
  All these indicates LVH with systolic strain.
- Poor R-wave progression : R amplitude is more in V2 than in V3 associated with Counterclockwise rotation
- fQRS (fragmentation) at V2-V3

Comments : The following reports with this case are also taken into consideration:

- ☐ Random blood sugar 250 mg % with HbA1c 8.2
- ☐ HRCT scan of thorax : Marked cardiomegaly with dilated main pulmonary artery , evidence of pulmoanry edema
- ☐ Cardiac Echo findings : Degenerative Aortic Valve Disease (Severe Aortic Stenosis with Moderate Aortic Regurgitration) , Mild MR
  Dilated LV with ejection fraction 25%
- ☐ NT-pro BNP 3903 pg/mL

❒ Degenerative severe aortic stenosis (AS) with moderate aortic regurgitation (AR) and functional mild mitral regurgitation (MR), aggravated by long-standing hyperglycemia.

- ○ AS is most likely calcific and degenerative, when associated with diabetes
- ○ AR is likely due to aortic root changes, possibly worsened by vascular stiffening from diabetes.
- ○ Mild MR appears functional, secondary to LV dilation and remodeling rather than primary valve disease.

- ❒ LVH pattern only with lead aVL, counterclockwise rotation, and severely reduced EF (25%) suggest LV stretching rather than true LVH.
    - ○ The lack of widespread LVH criteria suggests ventricular remodeling due to volume overload & dysfunction, rather than classical concentric hypertrophy.
    - ○ Counterclockwise rotation : This suggest the heart is rotating opposite to normal , often due to leftward displacement or dilatation of the right ventricle.
- ❒ Diabetic Cardiomyopathy (DCMP) with heart failure with reduced EF (25%) interacting with degenerative valvular AS/AR is likely responsible for LVH being limited to aVL.
    - ○ Diabetic cardiomyopathy contributes to LV dysfunction & fibrosis, further influencing the remodeling pattern seen on ECG.
    - ○ The combination of severe AS and heart failure (with pulmonary oedema) explains why LVH is not more widespread on ECG, as the ventricle is more stretched than truly hypertrophied.

NB : There is no finding on ECG suggestive of pulmonary hypertension as revealed by HRCT of thorax but this is equally true that in the presence of diffuse DCMP , the ECG evidence of pulmonary hypertension would be lacking.

Final impression :

- ☐ Diabetic cardiomyopathy (DCMP)
- ☐ Degenerative Aortic Stenosis with Aortic Regurgitation (with functional MR) , aggravated by coexisting DM2.

6. Take-Home Message

- ☐ In dilated cardiomyopathy (DCMP), the left ventricle (LV) enlarges and its contractile function declines. Despite this dilatation, the myocardium often exhibits hypertrophy. This form of hypertrophy differs from that observed with pressure overload conditions such as hypertension or aortic stenosis.
- ☐ LVH in DCMP is an important marker of disease severity and remodeling. While it results primarily from chronic volume overload and neurohormonal activation, additional factors like fibrosis, conduction abnormalities, and genetic predisposition contribute to its presence.
- ☐ Pathophysiology of LVH in DCMP
    - Ventricular overload leading to LV dilatation , initially compensated by ventricular stretching as per **Frank-Starling law** (perceived as pseudo LVH on ECG)
    - Neurohormonal Activation (RAAS & Sympathetic Overdrive)
        - ○ Myocardial hypertrophy as an adaptive mechanism
        - ○ Fibrosis , which contributes to electrical remodelling
- ☐ In DCMP , the heart exhibits eccentric hypertrophy .
  The pressure overload conditions such as hypertension or aortic stenosis exhibit concentric hypertrophy

- ECG findings in DCMP is having lower volatage criteria with poor R wave progression (PRWP) , late QRS transition at V5-V6 and clockwise rotation. The presence of LVH in DCMP correlates with worse prognosis and increased arrhythmic risk.
- Key Takeaways
  - Concentric LVH → High voltage, LV strain pattern, normal QRS duration
  - Eccentric LVH → Normal/low voltage, PRWP, QRS widening, conduction delays
- ECG Features in Diabetic DCMP + Aortic Stenosis
  - If DCMP is dominant → ECG resembles eccentric LVH (PRWP, low voltage, LBBB)
  - If AS is dominant → ECG resembles concentric LVH (high voltage, LV strain, normal QRS duration)
- Supplementary assessment, including imaging modalities like echocardiography or cardiac MRI, is highly recommended for precise evaluation.
- Patients with DCMP and LVH may respond differently to heart failure therapies. Regression of LVH with optimal therapy (ACE inhibitors, beta-blockers, and MRAs) correlates with better prognosis.
  Persistent LVH despite medical therapy may indicate poor response to reverse remodeling, necessitating CRT or ICD therapy in high-risk cases.

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DIABETIC CARDIOMYOPATHY AND ECG CHANGES : AN EMERGING CONCEPT

ECG

DIABETIC CARDIOMYOPATHY AND ECG CHANGES : AN EMERGING CONCEPT

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OUTLINE

Introduction
As Diabetic cardiomyopathy has become an increasingly recognized clinical entity among clinicians ; its better understanding is a 'must' for early diagnosis and the implementation of treatment strategies accordingly

Pathological-cum-clinical classification
Stage 1 : Early (Preclinical)
Stage 2 : Intermediate (Compensated Phase) Fibrosis increases →Stiff LV, Worsening diastolic dysfunction
Stage 3 : Advanced (Dilated , Failing Heart)

The pathological footsteps as correlative with ECG changes
Concentric hypertrophy → creeping fibrosis → LV dilatation with residual hypertrophy
Associated microvascular involvement
✓How to assess these findings on ECG ?

Illustration by ECG (An interesting case)

Take Home Message

References

Diabetic Cardiomyopathy and ECG changes : An emerging concept

A Narrative Review

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When the heart is hurt by its associated accompanion , it is broken down with a grieved cut – it will heal , but there will always be a scar. This tormentation does not fly away on the wings of time but rather deepens more. The shattering of a heart when being broken may remain tearful till the end.

The associated accompanion diabetes mellitus does not sweeten the heart with glucose-dependent energy but rather shifts the metabolic pathway towards a bitter turn.

- ☐ **Insulin resistance , a hallmark of Type 2 diabetes leads to inadequate glucose uptake by cardiomyocytes , resulting in energy deprivation and metabolic turmoil leading to a cascade of anatomical and functional remodelling of the heart.**
- ☐ **Diabetes mellitus in early stage expresses itself as HFpEF , while advanced cases expresses itself as HFrEF.**

ECG may prove itself the storyteller of the diabetic heart ; it deciphers the concerned patterns even before the heart succumbs to failure. The heart expresses its agony through the flickering signals on ECG.

1. Introduction (Keypoints)

- As Diabetic cardiomyopathy has become an increasingly recognized clinical entity among clinicians ; its better understanding is a 'must' for early diagnosis and the implementation of treatment strategies accordingly.
- This is a unique but distinct form of cardiomyopathy that occurs in patients with diabetes mellitus (DM), independent of other heart diseases like hypertension , any valvular or coronary artery disease (CAD). It is characterized by a combination of both diastolic and systolic dysfunction.
- It was first detected and described in 1972 by Rubler et al. when postmortem pathological reports revealed its nature in 4 diabetic patients who presented as heart failure without evidence of any concomitant pathology like coronary artery or valvular disease or even hypertension.
- Truly to say , diabetic cardiomyopathy looks to be an entity with shifting of energy substrate utilization – from a more balanced use of glucose to more reliance on fatty acid oxidation but this metabolic pathway is not so efficient so as to compensate the energy deficit in a diabetic patient. There are multiple factors as well involved in the pathogenesis of diabetic cardiomyopathy.

- It is manifested mainly as two subgroups :
  - Diastolic dysfunction (HFpEF-like phase) – Common in early diabetic cardiomyopathy ; ventricular hypertrophy preserves the myocardial function.
  - Systolic dysfunction (HFrEF-like phase) – ventricular dilatation develops as remodeling progresses.

  NB : Mixed DCMP phenotype (dilated with hypertrophy) – seen in advanced stages, with a combination of fibrosis, chamber dilation, and residual hypertrophy.

- Sex Differences in Diabetic Cardiomyopathy
  - Males with diabetes are more prone to develop systolic dysfunction (HFrEF)
  - Females with diabetes more commonly develop diastolic dysfunction (HFpEF) with preserved ejection fraction but in association with higher heart failure related mortality.
  - Estrogen's Protective Role: Females tend to have better mitochondrial function and endothelial response to diabetes compared to males.

- The ECG is very easily available tool which may bring the story of the diabetic heart on the surface – deciphers its pattern and whispers to the ears of clinicians – the different footsteps the heart meets with. Cardiac echo adds to its evaluation.

2. Pathological-cum-clinical classification

The pathophysiological spectrum in diabetic cardiomyopathy is a chain of events in continuum , which may be witnessed with its accompanying clinical scenario :

Stage 1 : Early (Preclinical)

Concentric hypertrophy → LV walls thicken due to hyperinsulinemia & AGEs (advanced glycation end-products).

Diastolic dysfunction appears first (HFpEF-like phase with EF ≥50%)

No major symptoms, but fatigue, mild dyspnea may be present.

Stage 2 : Intermediate (Compensated Phase)Fibrosis increases → Stiff LV, worsening diastolic dysfunction.

Microvascular dysfunction → Reduced myocardial perfusion. Subtle systolic dysfunction begins (mid-range EF 41-49%)

Arrhythmias (atrial fibrillation, QT prolongation) become common.

Stage 3 : Advanced (Dilated, Failing Heart)

LV dilates, leading to mixed remodeling (DCMP-like features).

Systolic function declines (HFrEF-like phase with EF<40%)

Heart failure symptoms (edema, pulmonary congestion, fatigue, exercise intolerance).

High risk of sudden cardiac death (SCD) due to ventricular arrhythmias.

NB :

- ☐ **Microvascular Disease**
  Capillary rarefaction (reduced density of small vessels) → Silent ischemia & impaired coronary flow reserve.
  Endothelial dysfunction → Reduced nitric oxide (NO) → Worse vasodilation & perfusion mismatch.

- ☐ **Autonomic Dysfunction & Arrhythmias**
  Diabetic neuropathy affects the heart → Reduced heart rate variability (HRV).
  Increased sympathetic tone → QT prolongation → Risk of sudden cardiac death (SCD).

3. The pathological footsteps as correlative with ECG changes

Here ECG changes can be well correlated with the pathological findings of diabetic cardiomyopathy , as enumerated below :

- Concentric hypertrophy → creeping fibrosis → LV dilatation with residual hypertrophy
- Other associated pathological changes
  - Microvascular involvement as reduction in density of small vessels with its concomitant endothelial dysfunction
  - Autonomic dysfunction with increased sympathetic tone

RV systolic and diastolic dysfunction

Type 1 DM and Type 2 DM have been found to be associated with RV dysfunction . This would be worthwhile to mention here that RV diastolic impairment seems to precede LV systolic dysfunction.

"Impairment of RV systolic function in patients with T2DM includes lower RV long-axis fractional shortening, reduced systolic excursion of tricuspid annulus, reduced pulmonary valve peak and mean velocities and impaired systolic strain. RV strain has also been correlated with levels of HBA1c. RV 3D echocardiography data in DM patients are limited. A study comparing 3D- and 2-D echocardiography showed that RV and right atrial strain were decreased in patients with pre-diabetes and DM as compared to controls. The early and deteriorating impact of diabetes on RV systolic and diastolic function develops independently from LV diastolic dysfunction, pulmonary hypertension and CAD."

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How to reveal ECG changes correlative with pathological footsteps

Concentric hypertrophy : (Increased LV wall thickness without significant dilatation)

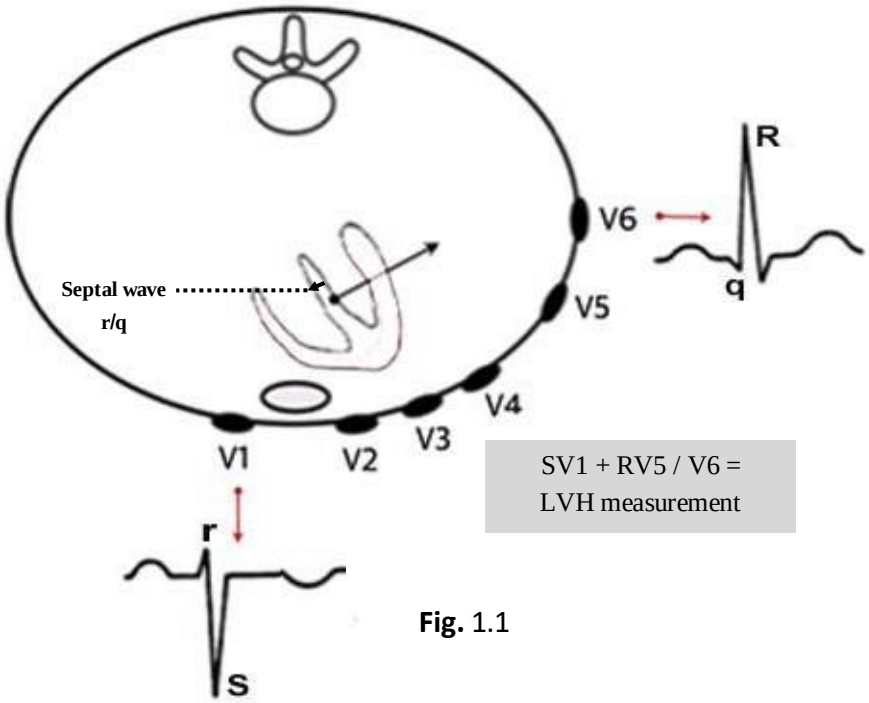


Fig. 1.1

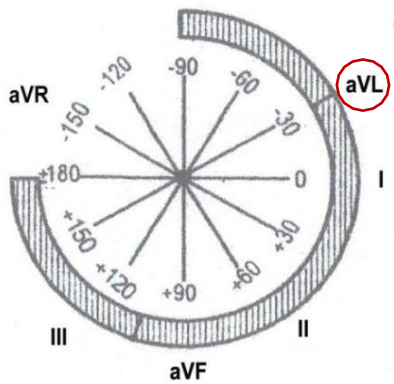


Fig. 1.2

LV dilatation with residual hypertrophy

- **Normally** , since septal wave is directed towards V1 **,** it inscribes 'r' as the first positive deflection in V1 and since septal wave is away from V6 , it inscribes q wave as a first negative deflection in V5/V6. And negative S in V1 and positive R in V6 both indicate the depolarization occuring through the L.Ventricle but at different thoracic levels.

- **Concentric hypertrophy :**
  The common criteria for left concentric hypertrophy in common use is
  Sokolow-Lyon criteria
  S-wave depth in V1 + tallest R wave height in V5 or V6 > 35 mm
  Sokolow-Lyon's index is the most used index, despite having the lowest sensitivity (20%) of all indexes. The specificity is high (>85%).
  (Other criteria like cornell-voltage criteria, Romhilt-Estes Point scoring system may also be used).

  R_{aVL} > 11 mm (with left axis deviation R in aVL should be >13 mm)

Supporting Non-voltage criteria :
Strain over lateral leads (I , aVL , V5, V6) : ST depression with T-inversion , caused by hypertrophy and related endocardial ischemia during repolarization phase.

- **Masked Hypertrophy :**
  Long -standing diabetic cardiomyopathy may present as low voltage in limb leads , inspite of hypertrophy (LVH paradox)

- **Assisted by imaging techniques**
  Echocardiography : confirms LV wall thickness , mass index , and systolic / diastolic dysfunction.
  Cardiac MRI (more sensitive than echocardiography in this context)

NB : Mild intraventricular conduction delay (IVCD) or left bundle branch block (LBBB) may be there due to subendocardial fibrosis and conduction slowing in diabetic cardiomyopathy.

Electrical axis assessment (Hexaxial lead system)

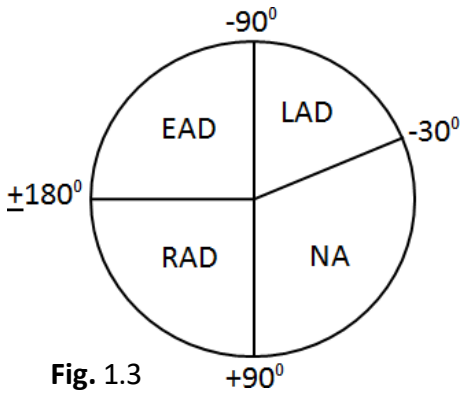


Fig. 1.3

- □ Normal Axis = -30^0 to $+90^0$
- □ Left Axis Deviation (LAD) = -30^0 to -90^0
- □ Right Axis Deviation (RAD) = $+90^0$ to $+180^0$
- □ North-West Deviation (NWD) This is also known as Extreme Axis Deviation = -90^0 to -180^0

Rule of 90^0

Any exploring lead placed within a range of 90^0 in respect to cardiac vector records positive deflection , at 90^0 equiphasic deflection or no deflection and beyond 90^0 negative deflection (with reference to hexaxial lead system)

- **QRS Vector with Concentric hypertrophy** Normal axis or Left axis deviation (LAD) due to thickened LV walls (the electrical vector may be shifted leftward between -30^0 to -90^0).

- **Possible QRS vector in masked hypertrophy** (LV dilatation with residual hypertrophy)
  - Normal to left axis deviation (LAD -30^0 to -90^0) – most common
  - **Indeterminate axis** : It is not equivalent to north-west axis. This terminology is used if QRS complexes are equiphasic in almost all the limb leads or if it cannot be plotted by the rule of 90^0.

    Mechanism :

    - Extensive myocardial fibrosis with disorganized depolarization pattern.
    - ✓ Loss of anterior septum (common in diabetic cardiomyopathy) can result in anterior infarction patterns, further altering the axis.
    - Biventricular enlargement can create conflicting electrical forces, leading to an indeterminate axis.

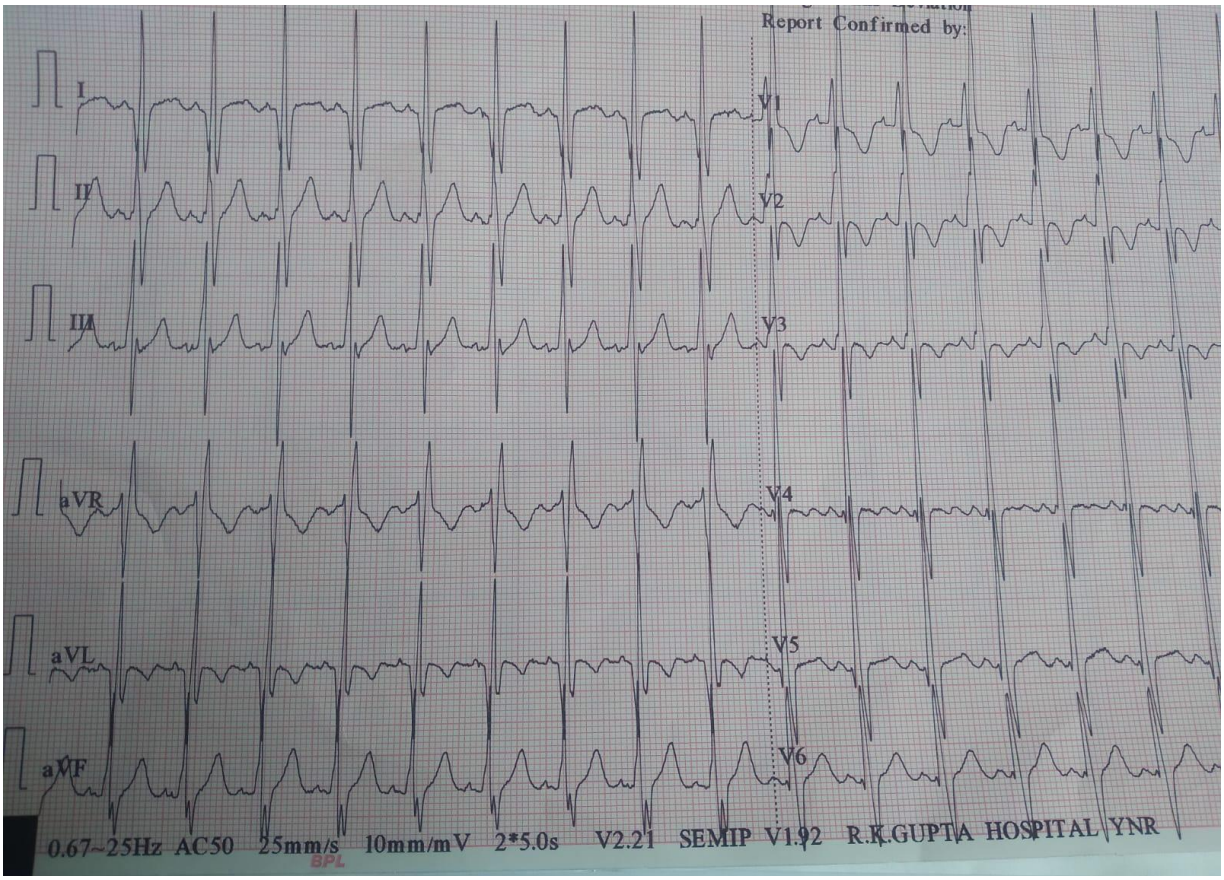
  - Right Axis Deviation (RAD) (+90° to +180°) – Less Common but Possible

    Mechanism :

    - Severe LV dilatation → secondary pulmonary hypertension → Right ventricular hypertrophy (RVH) → RAD
    - Independent RVH

4. Illustration by ECG

History : 60 yrs diabetic female presented with acute breathlessness , managed as acute LVF and B/L bronchoneumonia (Angiography could not be done because of concomitant pulmonary infections).



Source : Global Heart Rhythm Forum , on 16.03.2025 by Dr. R.K Gupta , Senior Consultant Physician , Yamunanagar , Haryana

Her echocardiography and X-ray Chest are posted below :

ECHOCRDIOGRAPHY :

- Left Ventricle Dilated, Concentric Left Ventricular Hypertrophy
- Regional Wall Motion Abnormally
- Anterior Wall, Anterior Septum Hypokinetic
- Severe Left Ventricular Systolic Dysfunction (LVEF 35%)
- Moderate Mitral Regurgitation , Mild TR , No PAH



Fig. 1.4

ECG findings	Comments
1. Indeterminate QRS axis (QRS complexes are equiphasic in so many limb leads and the axis cannot be determined by the rule of 90^0)	This may be due to the structural damage (fibrosis ± ischemic changes) to the interventricular septum leading to marked conduction defect therein. (anterior septum hypokinesia on cardiac echo also points towards the same)

Contd.

ECG findings	Comments
2. qR in leads I and aVL (Initial q is having much depth with R>11 mm)	Left ventricular hypertrophy with strain / ? old high lateral MI (deep q waves in lateral leads might be due to fibrotic tissue unable to depolarize)
3. RBBB-like pattern in leads V1-V3	Abnormal conduction through the fibrosed interventricular septum
4. R in V5 > 26 mm Total QRS voltage in all the 12-leads >175 mm	Left Ventricular hypertrophy
5. Sinus rhythm with ventricular rate > 100 bpm	Sinus tachycardia

Comments :

Based on the combined findings of ECG and Echocardiography

- The presence of indeterminate axis with RBBB like pattern in leads V1-V3 is most likely due to fibrosis creeping over the anterior interventricular septum (also evident on cardiac echo by the presence of anterior septum hypokinesia). All these indicate abnormal conduction through the fibrosed / ischemic interventricular septum.
- ECG and cardiac echo both demonstrate left ventricular hypertrophy
- The pattern of qR may indicate old high lateral MI , the amplitude of R>11 mm in lead aVL also indicates the left ventricular hypertrophy in association.
- On cardiac echo , the presence of moderate mitral regurgitation is possible with left ventricular dilatation.

In nutshell , ECG plus cardiac echo findings are suggestive of **advanced stage of diabetic cardiomyopathy.**

- Left ventricular dilatation , fibrosis plus concomitant concentric LVH
- Severe left ventricular systolic dysfunction (LVEF 35% , <40%)

(The patient presented with acute breathlessness which might be due to the combined impact of declined systolic function plus associated bilateral broncopneumonic changes)

Final impression

Diabetic cardiomyopathy with severe Left Ventricular Systolic dysfunction (LVEF 35%) associated with ? bilateral bronchopulmonary infection

5. Take-Home Message

- Diabetic cardiomyopathy is a distinct entity that occurs in patients with diabetes mellitus (DM), independent of other heart diseases like hypertension , any valvular or coronary artery disease (CAD). It is characterized by a combination of both diastolic and systolic dysfunction.

- This looks to be an entity with shifting of energy substrate utilization – from a more balanced use of glucose to more reliance on fatty acid oxidation but this metabolic pathway is not so efficient so as to compensate the energy deficit in a diabetic patient. There are multiple factors as well involved in the pathogenesis of diabetic cardiomyopathy.

- Diabetic-induced cardiomyopathy is an underdiagnosed but clinically significant cardiovascular complication. It begins as subclinical diastolic dysfunction which progresses to overt heart failure if not tackled early.

- Recent research suggests that diabetic cardiomyopathy is not a single entity but may have distinct subtypes based on structural abnormalities , assessed also with LVEF index on cardiac echo.
  - Diastolic dysfunction (HFpEF-like phase) – Common in early diabetic cardiomyopathy ; ventricular hypertrophy preserves the myocardial function.
  - Systolic dysfunction (HFrEF-like phase) – ventricular dilatation develops as remodeling progresses.
  - Mixed DCMP phenotype (dilated with hypertrophy) – seen in advanced stages, with a combination of fibrosis, chamber dilation, and residual hypertrophy.
  - Other associated findings

    ⇒ Microvascular involvement as reduction in density of small vessels with its concomitant endothelial dysfunction

    ⇒ Autonomic dysfunction with increased sympathetic tone

In nutshell , Diabetic cardiomyopathy is a specific cardiac manifestation of patients with diabetes characterized by left ventricular hypertrophy and diastolic dysfunction in the early phase and later on overt heart failure with reduced systolic function.
The pathogenesis of this condition is multifactorial , being recognised as a specific entity in the presence of insulin resistance and hyperglycemia.
NB : In Type 2 diabetes, high circulating insulin levels act as a potent growth factor. Insulin and insulin-like growth factor-1 (IGF-1) stimulate specific pathways that directly promote the growth of individual cardiomyocytes in thickness rather than length. Diabetes is frequently linked with arterial stiffness and at times hypertension , all these create a “Pressure overload” state , forcing the heart to thicken. Other factors like ‘Advanced Glycation End-products (AGEs)’ are also claimed to be responsible for concentric LVH by creating cross-link collagen in the heart , making the walls stiff and thick rather than dilated.

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HYPERTROPHIC CARDIOMYOPATHY : FUNDAMENTALS AND THE DUAL VECTOR CONCEPT

ECG

HYPERTROPHIC CARDIOMYOPATHY : FUNDAMENTALS AND THE DUAL VECTOR CONCEPT

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OUTLINE

Introduction
This is fundamentally a heredofamilial disease of sarcomeric dysfunction, where disordered contractile proteins lead to hypertrophy without proportional efficiency.

Pathophysiology in HCM
Myocardial disarray is a hallmark histopathological feature of hypertrophic cardiomyopathy .

Mechanistic principle in HCM based on dual vector concept
Hypertrophic cardiomyopathy is not only a structural and sarcomeric disease , but also a vectorial disease on ECG , where the pattern of depolarization and repolarization can be understood better to a dual vector framed work.

The applicability of the dual vector concept

- Isolated septal hypertrophy
- Apical and paraseptal hypertrophy
- Free left wall hypertrophy
- Symmetrical hypertrophic (diffuse) variant of HCM
- Right ventricular involvement (rare)

ECG changes in HCM other than those of segmental left ventricular hypertrophy

Illustration by ECG

Take-Home Message

References

Hypertrophic Cardiomyopathy : Fundamentals and the dual vector concept

A Narrative Review

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In some hearts, the past is not memory only—it is structure, carried silently across generations. Genes write their quiet script within the myocardium, long before symptoms find their utternaces through the suffering of the patient.

Hypertrophic cardiomyopathy is a discordant hymn of heredofamilial invocation, having inappropriate and asymmetrical myocardial hypertrophy by its morphemic expression without having an obvious inciting hypertrophy force –

A Flame of this Mystic Fire is burning within the heart.

- ☐ **This is fundamentally a disease of sarcomeric dysfunction, where disordered contractile proteins lead to hypertrophy without proportional efficiency.**
- ☐ **An unusual cluster of myocyte disarray at atypical ventricular sites creates a hypertrophic pattern on the ECG. This myocyte disarray, along with interstitial fibrosis, also serves as a substrate for arrhythmogenesis in hypertrophic cardiomyopathy.**

In heredofamilial cardiac disorder, the clinician must listen not only to the patient's heart, but to the family history as well that may voice aloud in favour of this chaos.

1. Introduction (Keypoints)

- Hypertrophic cardiomyopathy (HCM) is a heredofamilial disorder that is typically inherited as an autosomal dominant mode with variable penetration and expression.
- This is mainly attributed to inappropriate and asymmetrical myocardial hypertrophy by its morphemic expression without having an obvious inciting hypertrophy force. This hypertrophy can occur in any region of left ventricle or occasionally even the right ventricle , but interventricular septum is being the frequent site of involvement , in nearly quarter of patients, they may have also left ventricular subarotic obstruction in addition. This is the reason that this entity was also nomenclatured as hypertrophic obstructive cardiomyopathy (HOCM). But since this sort of obstruction is not always met with this disorder , this terminology had been replaced by HCM (Hypertrophic cardiomyopathy).
- Mutations involving genes coding for sarcomeric proteins are responsible for maximum number of cases.
- The electrocardiogram (ECG) is a vital, cost-effective, and highly sensitive (~90–95%) initial screening tool for Hypertrophic Cardiomyopathy (HCM), with over 90% of patients presenting abnormal findings. It is essential for identifying early, often asymptomatic disease—sometimes preceding echocardiographic evidence.

- **The most frequent abnormality on ECG is found to have an atypical LVH pattern with or without the abnormal Q waves.**

2. Pathophysiology in HCM

☐ In hypertrophic cardiomyopathy, sarcomeric mutations lead to hyperactive but energetically inefficient actin–myosin interactions, resulting in increased contractile stress, compensatory myocyte hypertrophy, and architectural disarray, ultimately producing a stiff and functionally impaired myocardium.

☐ The entire concept can be summarized as follows :

- Sarcomeric Gene Mutation → Abnormal sarcomere function.
- Hypercontractility (Early Phase) : Myosin–actin interaction becomes more active / prolonged resulting in increased contractile force initially with enhanced ATP consumptions.
- Subsequently energy utilization becomes inefficient due to associated poor energy fuel (ATP) → more work , less efficiency.
- Compensatory Hypertrophy → Myocytes enlarge
- Myocyte Disarray + Fibrosis
  Myocardial disarray is a hallmark histopathological feature of hypertrophic cardiomyopathy (HCM), characterized by a chaotic, non-parallel arrangement of cardiac muscle cells, often forming whorls or swirling patterns, in contrast to the orderly, parallel orientation of normal myocardial fibers. In HCM, these myocytes are often hypertrophied—meaning they are significantly wider and thicker than normal—and exhibit enlarged, sometimes bizarrely shaped nuclei.

Myocyte Disarray + Fibrosis → Arrhythmogenic foci with diastolic dysfunction (stiff ventricle with poor relaxation).

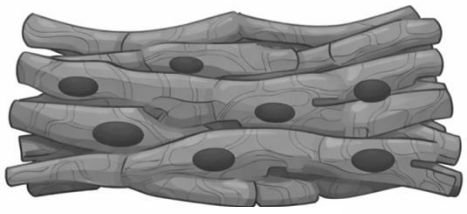


Fig. 1.1 **A**

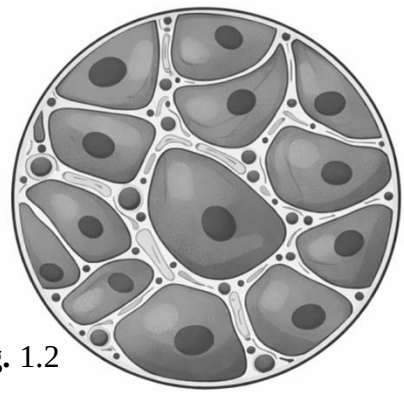


Fig. 1.2 **B**

A Orderly , parallel orientation of normal myocardial fibers

B Chaotic , non-parallel arrangement of hypertrophied myocytes (myocyte disarray)

3. Mehanistic principle in HCM based on dual vector concept

Hypertrophic cardiomyopathy is not only a structural and sarcomeric disease , but also a vectorial disease on ECG , where the pattern of depolarization and repolarization can be understood better to a dual vector framed work.

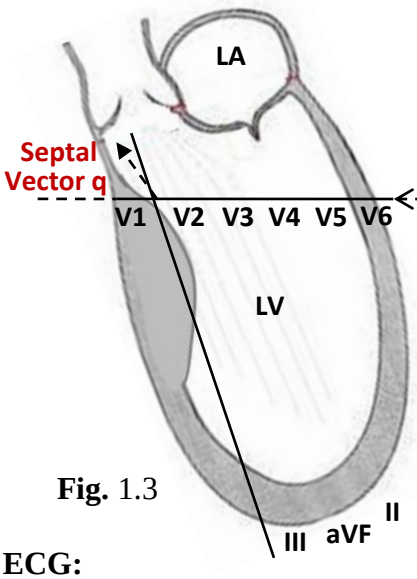
- **Dual vector concept :** The ECG in hypertrophic cardiomyopathy is best understood as a time-sequenced expression of two principal depolarization vectors: the initial septal vector, occurring within the first ~0.04 second, and the subsequent dominant vector generated by the hypertrophied myocardial mass in the affected region of the left ventricle.

  The final morphologic appearance of the ECG in HCM depends on the relative contribution and directional dominance of these vectors, which is determined by the site and extent of hypertrophy.

- **T-wave** denotes the phase of repolarization, which is influenced by heterogeneous recovery of the hypertrophied and disarrayed myocardium, and therefore , it often reveals itself by the associated deep symmetrical T-wave inversion.

4. The applicability of the dual vector concept

First illustrated in the setting of **isolated septal hypertrophy**, a recognized morphological variant of HCM. The term "isolated septal hypertrophy" refers to the predominant hypertrophic thickening of cardiac myocytes localized to the proximal interventricular septum.



(For simplicity the arrangement of leads V1 -V6 is illustrated horizontally , not placed so in real practice)

Fig. 1.3

Overall Impact on ECG:

i. Since the initial septal depolarization vector is directed predominantly from left to right (towards lead V1), it inscribes a relatively prominent R wave in V1.

ii. The lateral leads (V5–V6) and inferior leads (II, III, aVF), which view the septum from the opposite direction, record the initial depolarization moving away from them; hence, they inscribe deep, narrow, "dagger-like" Q waves in these leads.

iii. Because the dominant electrical forces are directed toward the hypertrophied interventricular septum (left → right), the contribution of the left ventricular free wall is relatively reduced, resulting in relatively smaller R-wave amplitudes in the remaining precordial leads.

Important Note:

It is important to note that the R-wave amplitude in the precordial leads is not significantly increased, indicating the absence of classical left ventricular free-wall hypertrophy. The presence of abnormal Q waves in the inferior and lateral leads, in the absence of LVH, reflects an augmented left-to-right septal depolarization vector, attributable to disproportionate septal hypertrophy.

Short Statement:

Dagger-like Q waves (in lateral and inferior leads) + prominent R in V1 + relatively small R waves in other precordial leads.

Diagnostic Clue:

The manifestation of abnormal Q waves in the absence of any evidence of left free-wall hypertrophy points toward dominant septal hypertrophy.

Electrocardiographic reflection of apical and paraseptal hypertrophy.

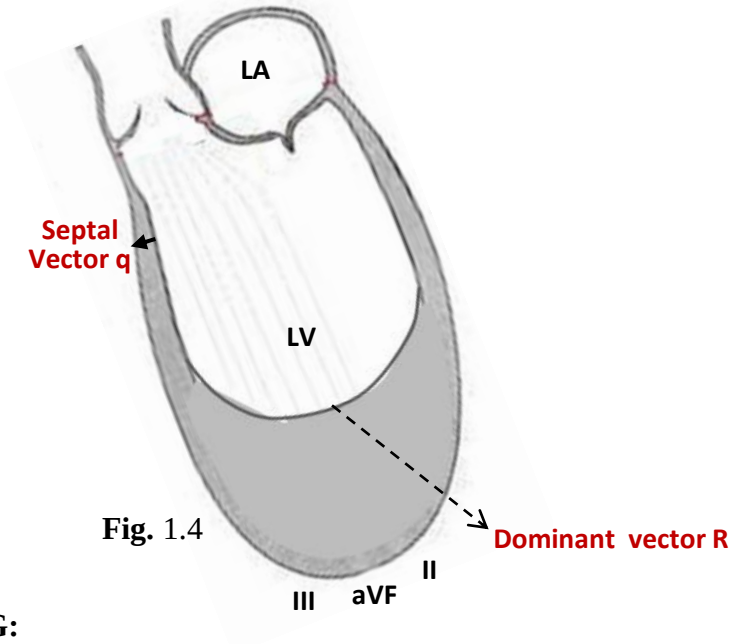


Fig. 1.4

Overall Impact on ECG:

i. The initial septal vector remains unaffected (non-hypertrophied zone), and therefore , physiological small q waves are preserved in the concerned leads.
ii. The dominant depolarization vector is directed toward the apical region, aligning with V4–V6 and inferior leads (particularly lead II), resulting in tall R waves in these leads.

iii. Repolarization is markedly abnormal due to apical hypertrophy with myocyte disarray, leading to deep, symmetrical, sharply pointed ("giant") T-wave inversions, most prominent in precordial and inferior leads.
iv. ST-segment depression is commonly present in the same leads, often appearing as a horizontal or downsloping "shelf-like" depression, continuous with the inverted T waves.

Short Statement:

Preserved initial q + dominant vector toward apex → tall R (V4–V6, II) + giant symmetric T-wave inversion (precordial & inferior leads).

Diagnostic Clue:

The combination of tall R waves with deep, symmetrical T-wave inversion in V4–V6 and inferior leads (maximal in lead II) is highly suggestive of apical HCM.

The electrocardiographic reflection of free left wall hypertrophy

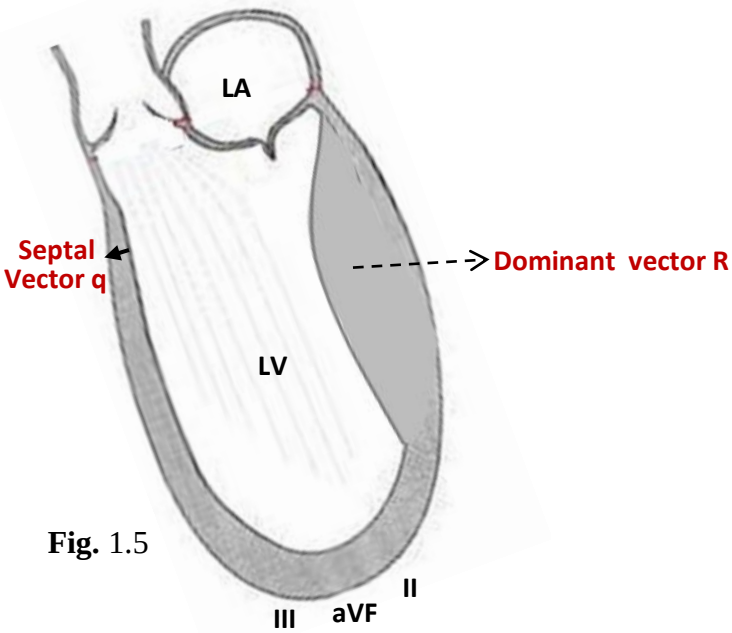


Fig. 1.5

Overall Impact on ECG:

i. The initial septal vector remains unaffected; therefore, physiological small q waves are usually preserved in the concerned leads.
ii. Hypertrophy of the left ventricular free wall is reflected by the classical ECG pattern of left ventricular hypertrophy (LVH) due to pressure (systolic) overload:
    Deep S waves in right-sided precordial leads (V1–V2)
    Tall R waves in left-sided precordial leads (V5–V6)
    The sum of S in V1 and R in V5 or V6 exceeds 35 mm (Sokolow–Lyon criterion)

iii. The repolarization abnormalities (strain pattern) are characterized by ST depression either horizontal or downsloping , most obvious in lateral leads (I, aVL , V5-V6) and deep T inversion in the same group of lateral leads. In contrast , T-wave is usually upright in V1 (systolic pressure overload is predominantly over lateral leads , not over V1).

Short Statement:

LVH assessment is viewed by Sokolow-Lyon criterion but the T-wave is upright in V1.

Diagnostic Clue:

If segmental hypertrophy is localized to the lateral free wall – specially when associated with deeply inverted T-waves in lateral leads (I , aVL , V5-V6), one should suspect this variant of HCM and let it be confirmed by further investigation like cardiac echo.

Electrocardiographic reflection of symmetrical hypertrophic (diffuse) variant of HCM

This diffuse ventricular wall thickening involves all segments of the left ventricle somewhat evenly

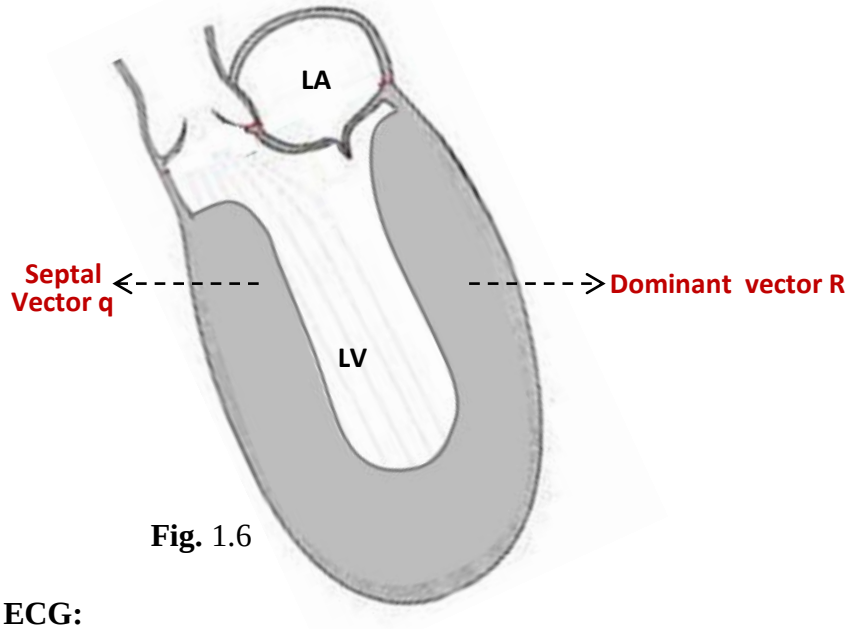


Fig. 1.6

Overall Impact on ECG:

i. Since hypertrophy involves the entire left ventricular (LV) wall rather than a localized segment, the ECG typically reflects increased overall QRS voltage (LVH) along with diffuse repolarization abnormalities.

ii. Large-amplitude QRS complexes are a hallmark, often fulfilling multiple voltage criteria for LVH.

iii. Repolarization abnormalities (“strain-like pattern”) include:
Widespread ST-segment depression, most prominent in lateral leads (V5, V6, I, aVL) and frequently extending to inferior leads (II, III, aVF)

T-wave inversion across multiple leads, usually deep but not of the "giant" (>10 mm) type, which is more characteristic of the apical variant of HCM.

iv. Deep, narrow ("dagger-like") Q waves may be present in lateral and inferior leads, reflecting abnormal septal activation with myocardial disarray.

Short Statement

Dagger-like Q waves + high-voltage QRS (LVH) + diffuse ST depression with deep T-wave inversion (lateral + inferior leads)

Diagnostic Clue

The presence of marked diffuse LVH criteria accompanied by widespread abnormalities, especially with dagger-like Q waves, strongly suggests a diffuse (symmetrical) hypertrophic cardiomyopathy phenotype.

ECG reflection of right ventricular involvement

This rare variant HCM involves the right ventricle, including the septum with one or more of the following findings :
Right axis deviation + right ventricular systolic overload + right atrial enlargement + complete or incomplete right bundle branch block.

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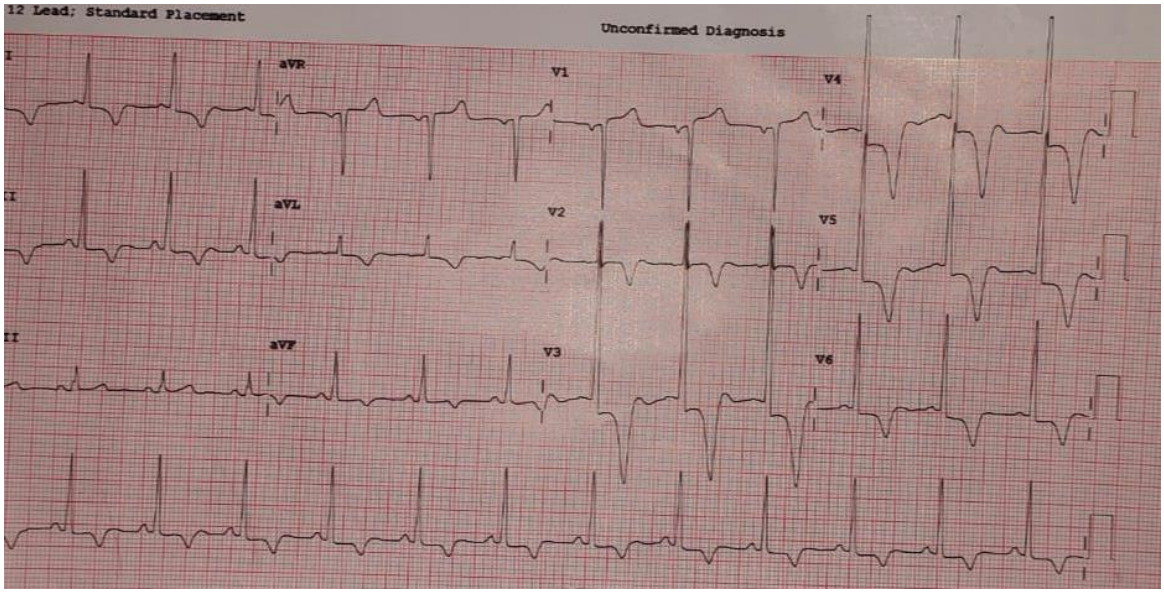
5. ECG changes in HCM other than those of segmental left ventricular hypertrophy

The electrocardiographic changes in association with segmental HCM have already been discussed in details. Now the other changes are listed below in brief.

- **Conduction defects**
  - Left anterior fascicular block – This is most significant manifestation of hypertrophic cardiomyopathy in children.
  - Left bundle branch block –incomplete LBBB is a frequent association of HCM , there is a disappearance of septal q wave in lead V5-6, lacking the evidence of deep /narrow q wave in this sort of block.
- **Left and/or right atrial abnormality** – Right atrial enlargement in the presence of left ventricular hypertrophy with typical ST and T changes is highly suggestive of hypertrophic cardiomyopathy.
- **WPW syndrome** – It is an uncommon association with HCM. Some authorities attribute delta like waves to incompete LBBB pattern due to pressure effect rather than WPW syndrome itself.
- **Short PR** occasionally seen , due to faster AV conduction or pre-excitation pathway
- At times **Prolongation of QT interval**
- **Arrhythmias** – commonly ventricular arrhythmias but supraventricular arrhythmias , may be seen even in asymptomatic individuals.

6. Illustration by ECG

A young male adult with dyspnoea on exertion



Source : Prof. Dr. A.N. Rai , Former Prof. & Head Medicine and Prinicipal ANMMCH , Gaya Bihar ; Chairman AIMS, Gaya

ECG findings :

- Evidence of left ventricular hypertrophy by Sokolow-Lyon criteria (SV1 + RV5 > 35 mm) in association with counterclockwise rotation from V2 – earlier appearance of R-wave.
- ST and T changes :
  There is also accompanying deep T-wave inversions with sharp pointed tips from V2-V4 (giant T-wave inversion in V3) PLUS the presence of ST depression in leads I , aVL , V2-V6.
  Important to note that T-wave in V1 is upright.
- There is totally absence of dagger-like Q wave throughout the tracings.
- Extra finding in association : Shortening of PR interval approx 0.10 second.

Discussion :

- The evidence of LVH by Sokolow-Lyon criteria in the absence of dagger-like Q wave is in favour of free left wall hypertrophic variant of HCM. Associated ST depression and T-wave inversion , as discussed above also support involvement of free left wall hypertrophy (the absence of dagger-like Q waves in throughout the tracings excludes possibility of diffuse symmetrical hypertrophy as a variant of HCM).
- It is to be noted that T-wave is upright in lead V1 , it is also a characteristic feature of free left wall involvement by hypertrophy (this positive T-wave is attributed to systolic pressure overload predominantly over lateral leads , not over V1).
- Shortening of PR interval without delta wave is in favour of LGL pattern in association or Fast AV conduction.

Impression based on ECG findings :

- Free left wall hypertrophic variant of HCM
- Shortening of QR interval , most possibly due to LGL syndrome / Fast AV conduction

(Cardiac echo also confirms this to be free left wall hypertrophic variant of HCM)

7. Take-Home Message

- Hypertrophic cardiomyopathy (HCM) is a heredofamilial disorder that is typically inherited as an autosomal dominant mode with variable penetration and expression.
- This is mainly attributed to inappropriate and asymmetrical myocardial hypertrophy by its morphemic expression without having an obvious inciting hypertrophy force. This hypertrophy can occur in any region of left ventricle or occasionally even the right ventricle.
- Pathophysiology in HCM : In hypertrophic cardiomyopathy, sarcomeric mutations lead to hyperactive but energetically inefficient actin–myosin interactions, resulting in increased contractile stress, compensatory myocyte hypertrophy, and architectural disarray, ultimately producing a stiff and functionally impaired myocardium.
- Myocyte Disarray + Fibrosis → Arrhythmogenic foci with diastolic dysfunction (stiff ventricle with poor relaxation).
- Hypertrophic cardiomyopathy is not only a structural and sarcomeric disease , but also a vectorial disease on ECG , where the pattern of depolarization and repolarization can be understood better to a dual vector framed work.
- **Dual vector concept :** The ECG in hypertrophic cardiomyopathy is best understood as a time-sequenced expression of two principal depolarization vectors: the initial septal vector, occurring within the first ~0.04 second, and the subsequent dominant vector generated by the hypertrophied myocardial mass in the affected region of the left ventricle.
  The final morphologic appearance of the ECG in HCM depends on the relative contribution and directional dominance of these vectors, which is determined by the site and extent of hypertrophy.
- Diagnostic clue as per dual vector concept :
  - The presence of dagger-like Q waves (usually in lateral and inferior leads) is suggestive of septal hypertrophy either isolated or as a part of diffuse symmetrical hypertrophy.
  - One has to see the precordial leads for the presence of added element of segmental LVH (usually in lateral /inferior leads).
- The classic "triad" of ECG changes in HCM includes left ventricular hypertrophy (voltage criteria , often with atypical pattern present in 50-70% of cases), deep "dagger-like" Q waves (20-50% of cases), and ST-T segment changes (ST-segment depression with deep, symmetrical, sharply pointed T-wave inversions) as the most prevalent abnormality, particularly in the lateral and inferior leads.
- ECG changes in HCM other than those of segmental left ventricular hypertrophy : Conduction defects (left anterior fascicular block , incomplete LBBB) , left and /or right atrial enlargement , WPW syndrome (?) , at times shortening of PR interval , prolongation of the QT interval , arrhythmias (ventricular and supraventricular arrhythmias).

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ISOLATED LVH IN aVL WITH CLOCKWISE ROTATION : A SUBTLE ECG CLUE TO HOCM

ECG

ISOLATED LVH IN aVL WITH CLOCKWISE ROTATION : A SUBTLE ECG CLUE TO HOCM

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Introduction

This article explores the phenomenon of isolated LVH in lead aVL with clockwise rotation on ECG as a possible early electrocardiographic marker of HOCM.

Why isolated LVH in aVL with clockwise rotation in HOCM ?

A. Isolated LVH in aVL (positioned at basal and high lateral LV wall) directing the QRS axis leftward and superiorly
B. Clockwise rotation : A clue to Posteriorly Directed Hypertrophy

A consideration to pertinent ECG changes as a subtle pointer to HOCM

The combination of isolated LVH in aVL with systolic overload and clockwise rotation suggests that the hypertrophy is affecting the basal septum and high lateral LV wall in a way that shifts electrical forces posteriorly.

Illustration by ECGs

Two interesting cases showing the pattern of 'isolated LVH in aVL with clockwise rotation' in favour of HOCM (confirmed on echocardiography)

Take Home Message

References

Isolated LVH in aVL with Clockwise Rotation : A Subtle ECG Clue to HOCM

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A scientific insight often emerges when one chooses not to follow the well-trodden path, but instead explores areas where only faint trails exist. While established norms make it easier to confirm typical findings, recognizing atypical presentations in medicine demands thinking beyond conventional frameworks.

With this perspective, one should avoid prematurely excluding a diagnosis on ECG, even when only subtle clues are present. Hypertrophic Obstructive Cardiomyopathy does not always manifest with striking electrocardiographic features; at times, it reveals itself only through minimal or isolated changes.e

- ☐ **A subtle clue, such as isolated left ventricular hypertrophy (LVH) in lead aVL, may provide an important diagnostic hint toward HOCM. Lead aVL reflects the high lateral wall of the left ventricle—an area that may be preferentially involved in certain phenotypic variants, including asymmetric septal hypertrophy.**
- ☐ **The diagnostic suspicion may be further supported by the presence of clockwise rotation. Structural remodeling, particularly involving the basal interventricular septum and adjacent high lateral wall, can alter the spatial orientation of ventricular depolarization, contributing to this pattern.**

Thus, ECG continues to serve as a dynamic and evolving diagnostic tool, capable of revealing nuanced insights even in conditions like HOCM—findings that should prompt further evaluation with advanced cardiac imaging

1. Introduction (Keypoints)

- ○ Hypertrophic cardiomyopathy usually encompasses an inappropriate hypertrophy of the myocardium occuring in the absence of any obvious cause like aortic stenosis or systemic hypertension and predominantly involving the interventricular septum in an asymmetrical manner with some accompanying clues on ECG , including dagger-like Q waves , high-voltage QRS complexes and deep T-wave inversions in the concerned leads , as per anatomical involvement of the myocardial territories.

- ○ **Certain cases of HOCM meet with diagnostic challenges in the absence of conventional patterns as just described.**

- ○ One such condition which is usually overlooked is an isolated LVH in lead aVL , particularly when associated with clockwise rotation in addition.

- ○ Recognizing such an atypical pattern with hypertrophic cardiomyopathy is an interesting topic to be reviewed through ECG.

- This study may explore the significance of high R-wave voltage in lead aVL as an indicator of LVH , associated with HOCM , specially if linked with clockwise rotation as an associate.
- Currently , there seems a paucity of established literatures specifically describing "isolated LVH in lead aVL with the associated clockwise rotation" as a distinct phenotype of HOCM. However , several studies and case reports have discussed other atypical ECG presentations with HOCM.

- Hypertrophic cardiomyopathy (HOCM) is a complex myocardial disorder often diagnosed based on echocardiographic findings of asymmetric left ventricular hypertrophy (LVH). However, early ECG recognition can be crucial, particularly in cases where classical ECG criteria for HOCM are absent. This article explores the phenomenon of isolated LVH in lead aVL with clockwise rotation on ECG as a possible early electrocardiographic marker of HOCM, needs to be confirmed by echocardiography.

2. Why isolated LVH in aVL with clockwise rotation in HOCM ?

This atypical pattern "isolated LVH in aVL with clockwise rotation" is a finding that, although subtle, may suggest underlying segmental hypertrophy and warrants further investigation with echocardiography.

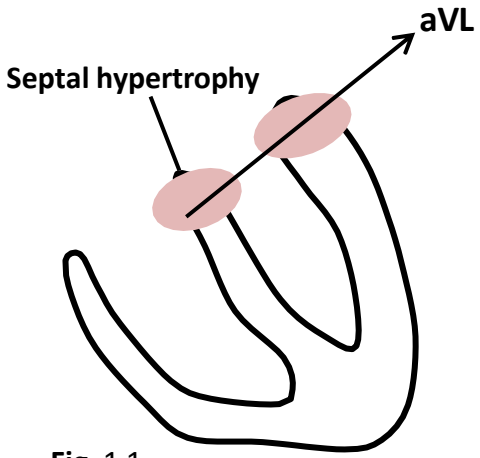


Fig. 1.1

A. Isolated LVH in aVL: A Unique ECG Signature

This is likely due to localized hypertrophy in the upper septal and corresponding upper lateral LV wall, directing the QRS axis leftward and superiorly

B. Clockwise Rotation: A Clue to Posteriorly Directed Hypertrophy

Clockwise rotation (delayed R-wave progression in precordial leads, with late transition in V5–V6) suggests posteriorly directed forces due to basal LV hypertrophy (more obvious if the hypertrophy is more localized to the basal and posterior LV segment rather than the septum)

3. A consideration to pertinent ECG changes as a subtle pointer to HOCM

LVH in aVL with clockwise rotation

Isolated LVH in lead aVL

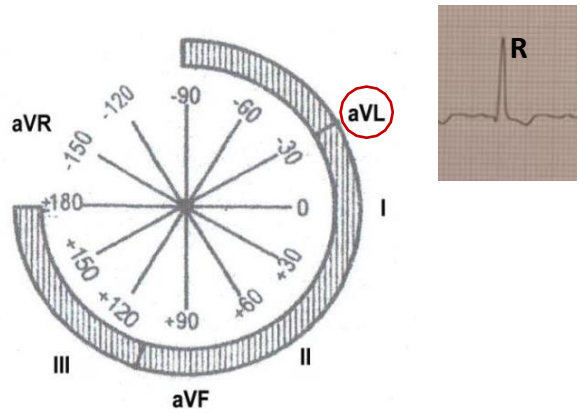


Fig. 1.2

- **Why aVL?**
  Lead aVL views the high lateral left ventricle—an area often preferentially hypertrophied in HOCM, especially in its asymmetric septal variant.
- R_{aVL} > 11 mm (with left axis deviation R in aVL should be >13 mm) may be an isolated but early marker of HOCM.
  This R may be attributed to increased afterload from left ventricular outflow tract (LVOT) obstruction.
- Strain over leads I and aVL :
  ST depression with T-inversion , caused by hypertrophy and related endocardial ischemia during repolarization phase.

Clockwise rotation

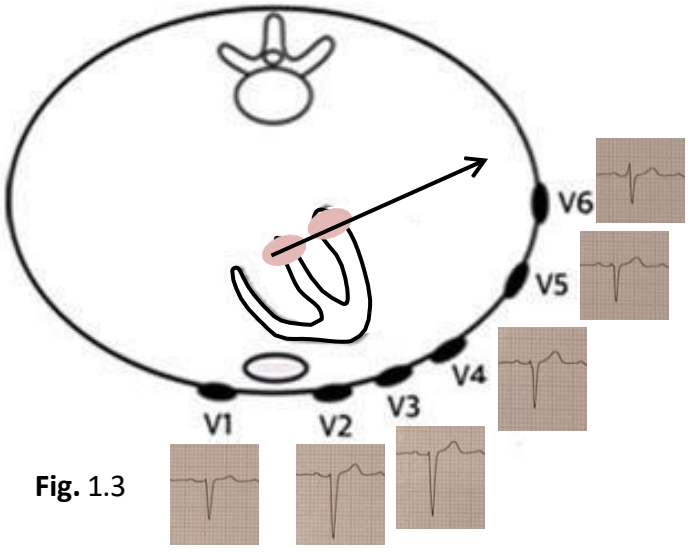


Fig. 1.3

- **Clockwise rotation**
  Normally, the R-wave progresses from V1 to V6, with the transition (R = S) occurring around V3 or V4. Here , in clockwise rotation, this transition shifts later (V5 or V6), indicating a relative posterior displacement of the involved segments.
- This may be attributed to :
  - ✓ Septal / high lateral LV wall hypertrophy pushing the heart posteriorly
  - ✓ Electrophysiological delay due to myocardial disarray (disorganised myocardial fibers)
  - ✓ Left ventricular outflow tract (LVOT) obstruction and its effect on ventricular geometry

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NB : The combination of isolated LVH in aVL with systolic overload, and clockwise rotation suggests that the hypertrophy is affecting the basal septum and high lateral LV wall in a way that shifts electrical forces posteriorly.

Be precautious of red signal



- ❒ When isolated LVH in aVL is seen with clockwise rotation on ECG , it is an alarming sign and HOCM should be strongly suspected
- ❒ This should prompt further echocardiographic evaluation, especially if there are symptoms like exertional dyspnea, chest pain , syncope, palpitation or a family history of sudden cardiac death.
- ❒ One should also auscultate the chest for the presence of mid-systolic murmur over the left parasternal area (ejection systolic murmur may be confused with valvular aortic stenosis)

Important clarification for some important findings on ECG in the present clinical scenario

- ➢ The absence of Q wave : The usual deep septal Q waves of HOCM may be absent, as hypertrophy in these cases does not predominantly involve the anteroseptal wall.
- ➢ Pseudo-Delta Waves : In some cases, initial slurring of the QRS complex may be observed, mimicking a pseudo-delta wave. This may result from fibrosis-related conduction delay in hypertrophied myocardial segments (prone to ventricular arrhythmias→SCD).
- ➢ T-Wave inversion may not be deep in this scenario due to the following consideration:

  In typical HOCM with severe asymmetric septal hypertrophy, we expect deep T-wave inversions in the concerned leads due to repolarization abnormalities caused by massive anteroseptal hypertrophy.However, when hypertrophy is distributed across both anterior and posterior segments at the basal/high lateral LV wall, the repolarization forces may partially cancel each other on the frontal plane.This neutralization effect can blunt the expected T-wave inversions, making them less prominent than in classical cases of HOCM.

The traditional reliance on global LVH patterns and deep T-wave inversions in precordial leads may cause certain cases of HOCM to go unnoticed. Recognizing the significance of isolated LVH in aVL with systolic overload , especially when accompanied by clockwise rotation , can help in unmasking HOCM at its earlier stages.

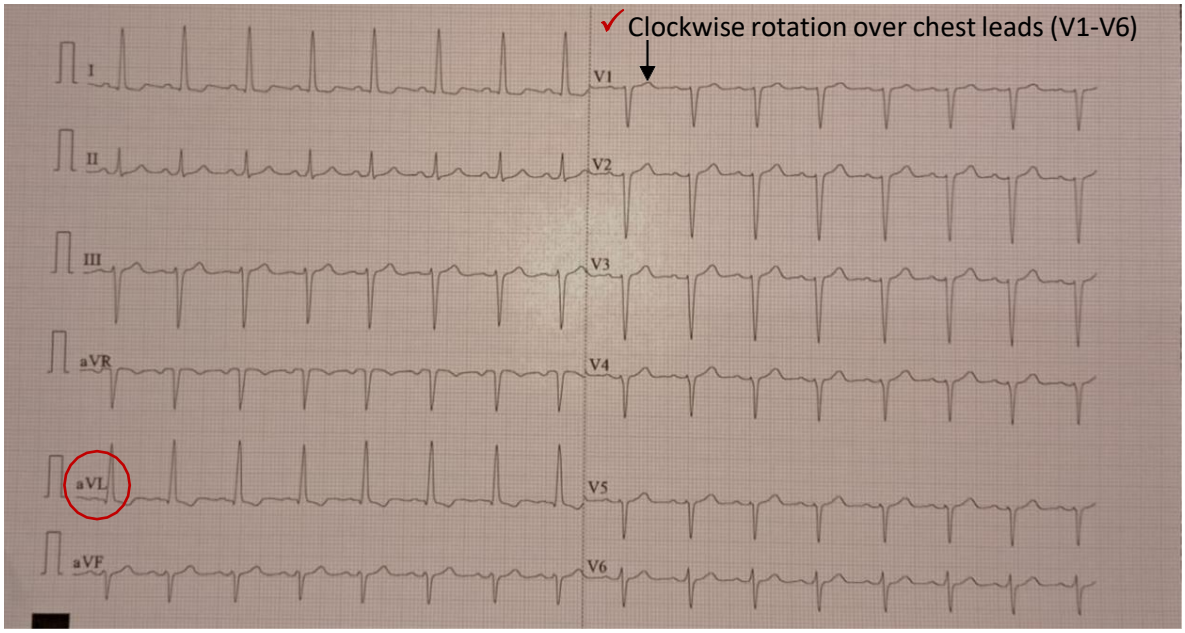
Assisted by imaging techniques

- **Echocardiography :** Echo is diagnostic of HOCM
  Asymmetrical septal hypertrophy (ASH) , systolic anterior motion (SAM) of the MV apparatus which may abut the IVS , mid-systolic AV closure and fluttering ,etc. And the continuous Doppler may show a peak velocity across the left ventricular outflow tract.
- **Cardiac MRI with late gadolinium enhancement (LGE)** to assess and quantify fibrosis.

4. Illustration by ECGs

Case No. 1

50 years aged female presented with 2 months h/o distension abdomen , Fever 2 months , Dyspnoea on exertion - NYHA IV (USG showed ascites , 4L of fluid drained from abdominal cavity – exudate in nature , ATT started) CVS Exam : systolic murmur over LPSA



Source : Global Heart Rhythm Forum on 20-03-2025 by Dr. R.K Gupta , Senior Consultant Physician , Yamunanagar , Haryana

❒ **Please review the following for proper understanding the concerned ECG changes:**

ECG findings	Comments
• **R in aVL>11 mm** (QRS axis +20 degree) , associated with ST depression / T-inversion in leads aVL and I)	Isolated LVH in lead aVL with systolic overload
• **Clockwise rotation over chest leads**	This indicates shifting of electrical forces posteriorly by the hypertrophied basal septum and high lateral LV wall
• Mild slurring at the beginning of the ascending limb of QRS complexes , obvious over leads II and V6	Possibly fibrosis –related conduction delay in hypertrophied myocardial segments (this subtle QRS slurring may be mistaken for pre-excitation - WPW). It is a risk factor for ventricular arrhythmias and sudden cardiac death (SCD) in HOCM.
• Accompanying sinus tachycardia	----

❒ **Cardiac echo : Salient features**

- Hypertrophic cardiomyopathy
- Peak LVOT gradient = 66 mmHg Significant LVOT obstruction +, SAM+
- Continuous Doppler showing a peak velocity across the left ventricular outflow tract
- Mild MR , mild AR ; No AS/TR

'This dynamic obstruction is more pronounced at the later stages of systole. As the LV becomes empty, LV cavity size is comparatively smaller and the anterior mitral leaflet moves anteriorly to abut against the septum. It may be present at rest or become more pronounced with valsalva maneuvre or exercise.

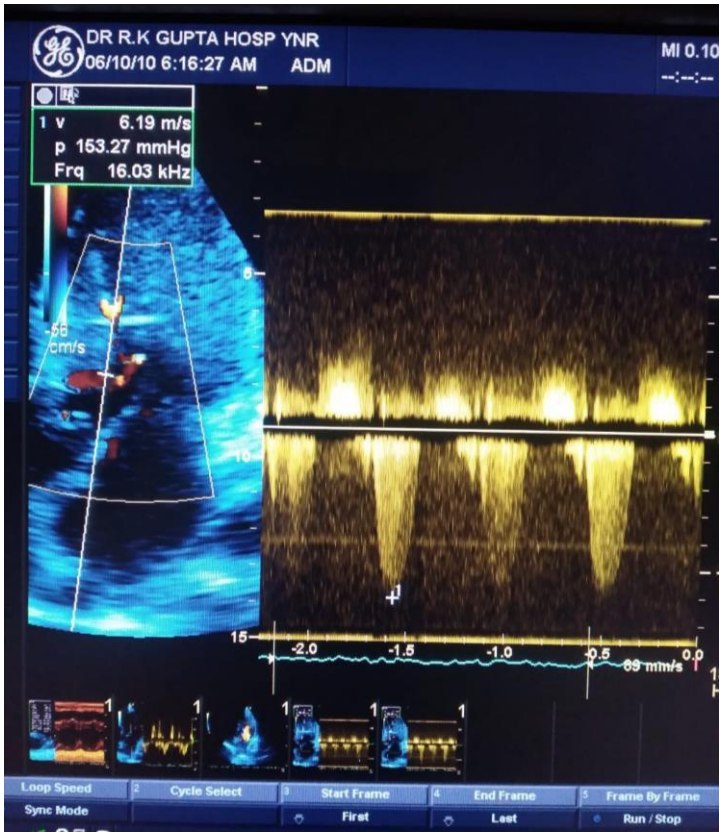


Fig. 1.4

NB : In some individuals with HOCM, the most dangerous period is at the end of vigorous exercise , e.g. during a race competition. At this time, ventricular volume diminishes as cardiac output and heart rate decrease , catecholamine drive is having a downgoing pattern, and there may be changes in circulating electrolyte concentrations , such as K+. These features all combine to increase the risk of syncope and sudden cardiac death by increasing the likelihood of increasing left ventricular outflow gradient and ventricular arrhythmias (**Ref :** ECHO MADE EASY , Second Edition by Sam Kaddoura, P 80-82**)**

❒ **On auscultation ejection systolic murmur heard over LPSA**

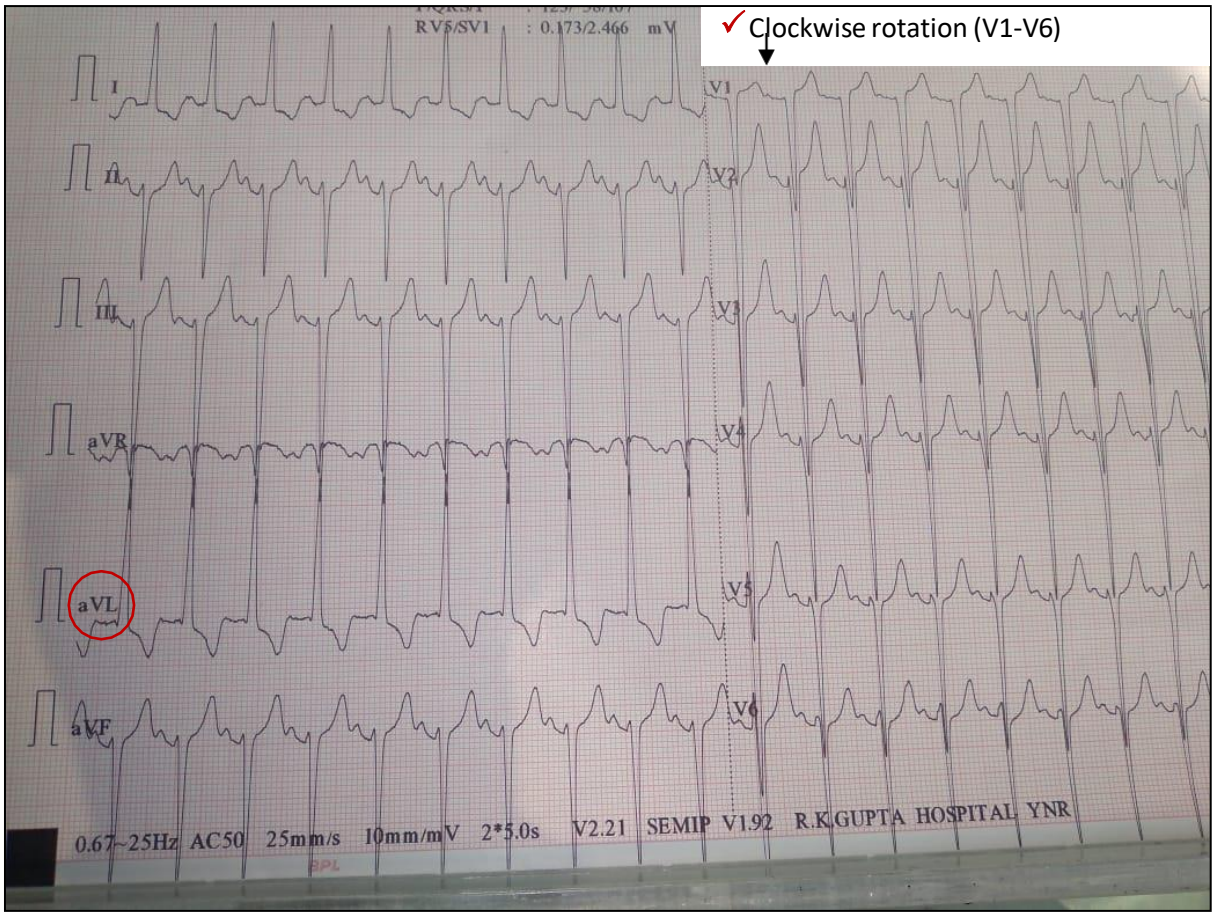
Comments :

- The combination of isolated LVH in aVL with systolic overload associated with clockwise rotation over the chest leads suggests hypertrophic cardiomyopathy.
- Cardiac echo findings , as discussed above are diagnostic of HOCM.
  - SAM is hallmark of HOCM – related LVOT obstruction.
  - Patient with peak LVOT gradients $\geq$ 50 mmHg are at higher risk for HOCM related symptoms.
- The presence of mid-systolic murmur over LPSA in association is also the pointer to the same diagnosis.

Case No. 2

59 years female , diabetic 20 years , hypertensive 7 years , came for medical fitness for surgery

BP 160/80 mmHg , hemodynamically stable no h/o chest pain or dyspnoea, systolic murmur over LPSA also heard.



Source : Global Heart Rhythm Forum on 24-01-2025 by Dr. R.K Gupta , Senior Consultant Physician , Yamunanagar , Haryana

❒ Please see the followings :

ECG findings	Comments
• **Marked R in aVL>13 mm** (left axis deviation , QRS axis -50^0) , associated with ST depression / T-inversion in leads aVL and I)	With left axis deviation R in aVL > 13 mm is the pointer of isolated LVH in lead aVL Associated systolic overload.
• **Clockwise rotation over chest leads with prominent upright T-waves**	This indicates shifting of electrical forces posteriorly by the hypertrophied basal septum and high lateral LV wall
• Accompanying sinus tachycardia	----

❒ **Cardiac echo : Salient features**

Hypertrophic Cardiomyopathy with mid - cavity gradient of 85 mm Hg

A "dagger-shaped" signal with CW

NB :

- **Mid-Cavity Obstruction** (MCO) : Unlike classical hypertrophic obstructive cardiomyopathy (HOCM) , here obstruction stands within the body of the LV.
- A gradient of 85 mmHg indicates severe obstruction
- **Dagger-Shaped CW Doppler's signal** suggest a progressive increase in velocity during systole , creating a late peaking , with dagger-like appearance ; this is considered a key diagnostic feature for obstructive HOCM

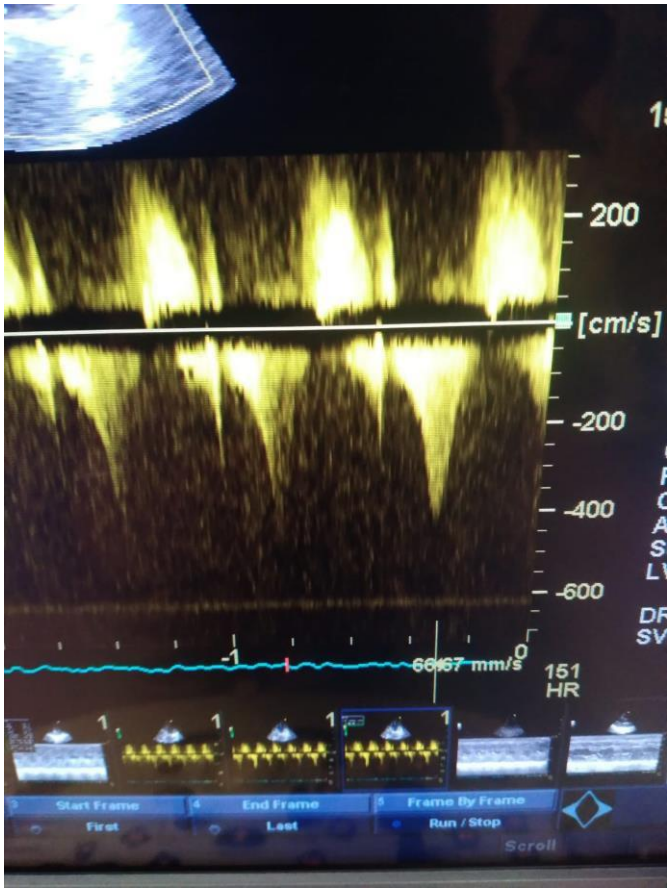


Fig. 1.5

❒ **On auscultation with ejection systolic murmur heard over LPSA**

Comments : All the followings are in favour of HOCM

- Isolated LVH in aVL in the presence of left axis deviation with systolic overload , linked with clockwise rotation over the chest leads
- Cardiac echo : Hypertrophic cardiomyopathy with cavity gradient of 85 mmHg and dagger-shaped signal on Continuous Wave Doppler.
- The presence of ejection-systolic murmur in association over LPSA is also the pointer to the same diagnosis.

5. Take Home Message

- The conventional reliance on global LVH patterns and deep T-wave inversions in precordial leads may cause certain cases of HOCM to go unnoticed. Recognizing the significance of isolated LVH in aVL, especially when accompanied by clockwise rotation and systolic overload can unmask HOCM at its earlier stages
- While LVH in aVL in association with clockwise rotation alone is not diagnostic of HOCM, in the right clinical context—especially with systolic overload—it may serve as an early or subtle ECG marker. Therefore , there is a dire need for vigilance in ECG interpretation , as early detection of HOCM can prevent complication like sudden cardiac death.
- Lead aVL, placed at –30 degree on the frontal plane, provides an anatomical view of the high lateral and basal septal regions of the left ventricle. In HOCM with asymmetric septal hypertrophy (ASH), these regions are often preferentially thickened, directing electrical forces towards aVL. As a result, the R-wave amplitude in aVL may become disproportionately prominent, even in the absence of significant precordial LVH findings.
- Clockwise rotation, detected by delayed R-wave progression in precordial leads (shifted transition to V5 or V6), is another subtle but crucial clue. The hypertrophied basal septum and high lateral LV wall in HOCM can shift the electrical axis posteriorly, delaying septal activation and altering QRS transition.
- In this context isolated LVH in aVL with clockwise rotation should prompt further imaging with echocardiography or cardiac MRI to confirm hypertrophy distribution and associated fibrosis if any (subtle QRS slurring on ECG may indicate underlying myocardial fibrosis).
- More case studies are required to validate the hypothesis – “Isolated LVH in aVL with clockwise rotation is a subtle ECG clue to the diagnosis of HOCM”.

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TAKOSUBO CARDIOMYOPATHY (TCMP) : A RUNNER THROUGH STRESS

ECG

TAKOTSUBO CARDIOMYOPATHY (TCMP) : A RUNNER THROUGH STRESS

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OUTLINE

Introduction

Takotsubo cardiomyopathy is a special type of cardiomyopathy that is characterized by a transient stress induced reversible dysfunctioning with ballooning of a portion of the left ventricle of the heart , most often over its apex.

Pathophysiology

- Catecholamine Surge
- Microvascular spasm
- LVOT obstruction

ECG changes associated with Takotsubo cardiomyopathy

- ST elevation
- T-wave inversion
- Q-wave
- QTc prolongation
- Cardiac arrhythmias

Differentiation of these ECG changes from STEMI

A diagnostic approach to TCMP

Illustration by ECG

Take Home message

References

Takosubo Cardiomyopathy (TCMP) : A runner through stress

A Narrative Review

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The Fire of stress is often said to be the slayer of our existence; yet, in truth, it is not stress itself that destroys us, but our response to it.

O Heart ! Thou art not an exception—the flame of stress may indeed unsettle thee with its anguish.

- **A sudden biochemical surge of catecholamines, instead of protecting, may transiently impair thy function.**
- Thy movements become disordered—dyskinetic at the apex, which balloons outward, and hyperkinetic at the base, at times contributing to dynamic left ventricular outflow tract (LVOT) obstruction.

This entire spectrum of cardiac changes is known as stress cardiomyopathy, wherein the underlying mechanism is believed to involve catecholamine excess leading to microvascular dysfunction, myocardial stunning, and transient myocyte injury.
By the grace of the Almighty, these changes are usually not permanent—recovery of ventricular function typically occurs within days to weeks.

(Also known as Takotsubo cardiomyopathy, apical ballooning syndrome, broken heart syndrome, or ampulla cardiomyopathy.)

1. Introduction

Takotsubo cardiomyopathy is a special type of cardiomyopathy that is characterized by a transient stress induced reversible dysfunctioning with ballooning of a portion of the left ventricle of the heart , most often over its apex. It was originally identified and described in Japan by Sato et al. in 1990. The word 'Takotsubo' was coined from the name of a pot used by Japanese fishermen to trap octopuses. The apical part of the heart changes its contour like a round pot with its narrow neck – very much similar to the octopus trapping pot.

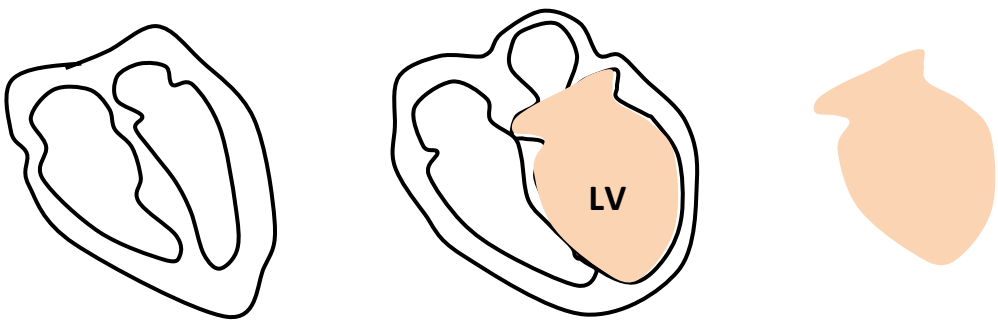


Fig. 1.1

In TCMP, apical dyskinesia with subsequent ballooning creates LV appearance remarkably similar to the octopus pot.

Since its recognition this condition was found to be prevalent all over the globe affecting any one with any ethnic background. However , more than 90% of the reported cases are in postmenopausal women , usually over 60 years, being devoid of oestrogen protection . There is also evidence that upto 5% of women being suspected of having a heart attack belong to this group of disorder. The most people recover rapidly within a few weeks without any trace of heart damage. The prevalence has been shown to be in between 1.2-2.2% of all the patients who presented with suspected acute coronary syndrome.

In the majority of cases , this illness is preceded by the events of either intense emotional stress or physical stress. The entire clinical scenario is attributed to catecholamine surge as a result of emotional or physical upset. It would be worthwhile to mention here that there is an absence of significant coronary artery disease in TCMP as accessed by angiograms (diameter stenosis <50%) or angiographic evidence of any acute plaque rupture.
TCMP resembles very much on ECG with acute ST elevation MI. This is a clinical dictum that with any patient having suspicion of acute coronary syndrome (ACS) , ECG recording should be the initial step. Under circumstances it would be a wiser step to have the basic concept of 'Takotsubo cardiomyopathy' to distinguish it from myocardial infarction and also allied conditions as well. It is a rule to consider the affected individual to have a heart attack until proven otherwise.

2. Pathophysiology

The underneath triad consisting of catecholamine surge , microvascular spasm and LVOT obstruction is mainly responsible for its causation.

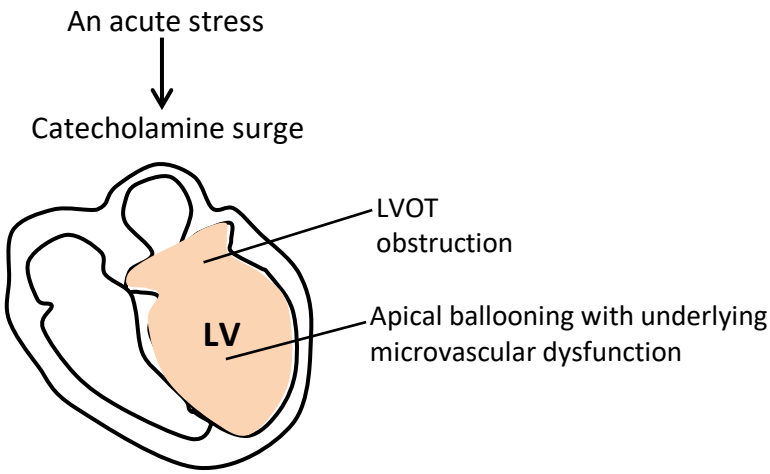


Fig. 1.2

❑ Catecholamine surge

The biochemical bursting with catecholamine surge seems to be the initiating factor. This is to be noted here that the apical part of the left ventricle has the highest density of sympathetic nervous system fibers , the activation of which by catecholamines causes microvascular spasm resulting in lesser blood supply to the apical zone. Not to surprise , this seems to be the

causation of regional wall dyskinesia , involving mainly the left ventricular apex with hyperkinesia of the basal left ventricular segment.

There are different types of wall motion abnormalities associated with 'Takotsubo cardiomyopathy' , as mentioned below :

- Classical type with apical ballooning (the most common)
- Reverse apical ballooning i.e. Hyperdynamic apex with akinesia at the base of the left ventricular wall.
- Mid ventricular type involving the mid left ventricular wall , sparing the base and apex.
- Localized type involving a segment of the left ventricle , usually the anterior wall.

The regional wall motion abnormalities appear first in the cardiac echography and changes in ECG appear approximately one day later.

❑ Microvascular spasm

The sequence of events are as follows :

Catecholamine surge ⟶ action on the apical sympathetic nervous system fibers ⟶ microvascular spasm with myocytes-dysfunctioning.

❑ LVOT obstruction

In 10%-50% cases of TCMP , there is an underlying some degree of left ventricular outflow tract (LVOT) obstruction. The causal factor to this obstruction is said to be the hyperkinesia of the basal segment of the left ventricle. This is a known fact that there is a declining coronary perfusion from the basal to the apical zone and therefore , when the apical zone is akinetic , the basal zone becomes hyperkinetic.

Two factors are to be mentioned here :

(i) **The basal hyperkinesia** narrows the systolic LVOT creating a narrow pathway and thus , promoting the 'venturi effect'. The Venturi effect is the reduction in fluid pressure that results when there is a flow through the constricted section. LVOT obstruction working as a narrowed outlet causes a drop in its blood flow pressure which may cause systolic anterior movement of the mitral valve as well with further increasing the obstruction.

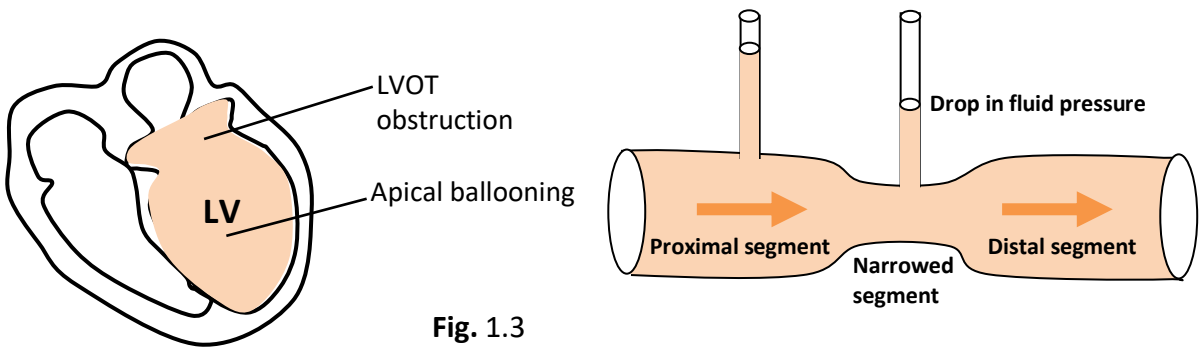


Fig. 1.3

(ii) **Pre-existing sigmoid-shaped septum** – a normal part of the aging process may also contribute to LV obstruction as well to some extent.

Most of the cases have full recovery with dissolution of this LVOT obstruction but it should be kept in mind that this LVOT obstruction is dynamic in nature with a poor clinical outcome with the heart. With LVOT obstruction there may be some serious complications , such as CCF or cardiogentic shock. The conventional therapy with ionotropes and / or vasopressures to combat the situation with cardiogenic shock may worsen LVOT obstruction and can be fatal. Therapy should rather be focused on reducing the LVOT gradient and does include the fluid administration to improve preload and beta-blocker therapy to increase diastolic filling time.

✓Complications are encountered in some patients , which may include the followings :

- Mild to moderate Mitral regurgitation
- Acute left heart failure
- Dynamic left ventricular outflow tract obstruction, which may lead to CCF or cardiogenic shock.
- LV mural thrombus
- Ventricular arrhythmias
- At times even rupture of the ventricular wall

3. ECG changes associated with Takotsubo cardiomyopathy

A consideration : **ECG changes are the reflection of underlying 'myocardial stunning' as a result of overwhelming catecholamine surge but the apical dyskinesia and basal hyperkinesia recover earlier within a few weeks**. At the level of cardiac myocytes there might be some biochemical abnormalities like abnormal energy utilization , production of oxygen free radicals , abnormal calcium influx associated with some auto-regulatory phenomenon. With the resolution of apical dyskinesia and basal hyperkinesia there is an earlier journey towards recovery from these deleterious effects within a few weeks.
The regional wall motion abnormalities appear first on the cardiac echo in most cases and changes in the ECG appear subsequently one day later.

The ECG findings are mainly recorded in precordial leads , at times even may be exteneded to limb leads.

ST elevation
During the initial stage of myocardial stunning there is a current of injury maximum over the subepicardium and thereby , the current flows from subendocardium towards epicardium with ST elevation. The ST elevation usually persists for 1 to 3 days i.e. till the resolution of this initial acute phase.

T-wave inversion
Dynamic and diffuse T-wave inversions are the most consistent ECG findings in Takotsubo cardiomyopathy. These T-wave changes are observed during the next continuing phase of myocardial stunning lasting for a few weeks. There is a reversal of

repolarization , now from endocardium instead of epicardium – inscribing the change as T-wave inversion.

ST and T changes occur mainly in precordial leads , more rarely in limb leads as well. The stay of ST segment elevation is shorter whereas that of T-wave inversion is longer. This is to be noted that there is no reciprocal changes due to the diffuse involvement of left ventricle in Takotsubo cardiomyopathy.

- **Q-wave**
  There may be the inscription of Q-wave due to associated apical left ventricular dyskinesia. The disappearance of Q-waves is assumed to be related to the recovery of myocardial motion abnormality without necrosis.

- **QTC prolongation** reported in a substantial proportion of patients with TCMP.

- **Cardiac arrhythmias**
  - Sinus tachycardia due to catecholamine surge
  - Atrial fibrillation is the most common form of arrhythmia despite the TCMP being of ventricular origin. It is again presumed to be due to increased catecholamine surge or associated myocytes-dysfunctioning.

These ECG changes are often dynamic and transient , and their presence or absence depends upon when ECGs are recorded after the symptoms onset. That's why , analysis of ECGs over time is considered to be helpful in the diagnosis and management of the patients with TCMP.

4. Differentiation of these ECG changes from STEMI

Takotsubo cardiomyopathy is very difficult to be distinguished from STEMI and no ECG criteria can be safely used to differentiate between these two conditions. However , the following points should be considered for the purpose :

- The amplitude of ST-segment elevation is smaller in patient with TCMP than in patient with STEMI.
- It would be worthwhile to mention here that reciprocal ST-segment changes are absent in TCMP.
- Usually the patient with TCMP presents with diffuse T-wave inversion in the background of emotional or physical turmoil.
- A retrospective case study by Ogura et al demonstrated that higher ST elevation in leads V4-V6 compared to V1-V3 with the absence of pathological Q-wave and reciprocal changes in the inferior leads showed high sensitivity and specificity to differentiate Takotsubo cardiomyopathy from anterior STEMI.

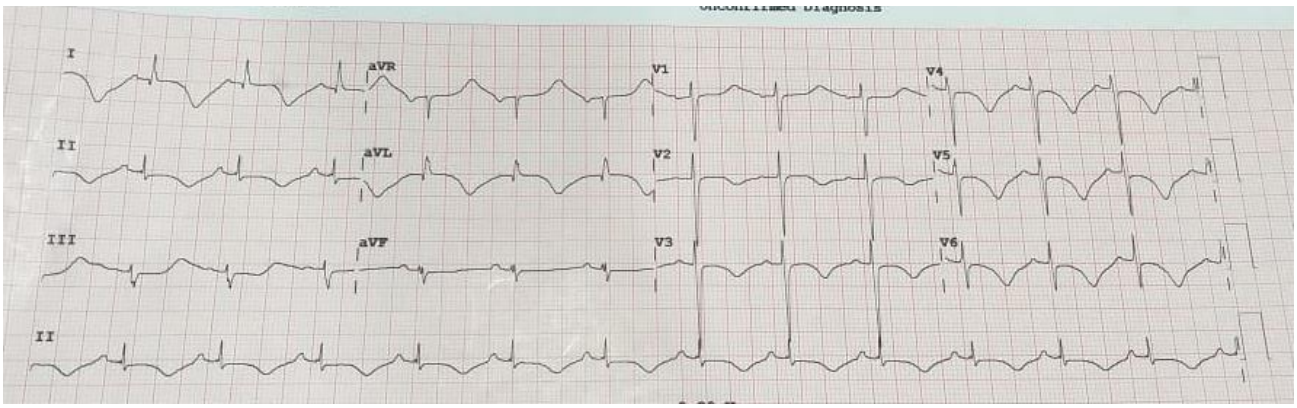
However , it is very difficult to distinguish TCMP from an ACS on the basis of ECG changes alone ; therefore , access to emergency Coronary angiography should never be delayed.

5. A diagnostic approach to TCMP

TCMP induces ischemic chest pain and ECG changes as described are associated with moderate elevation of cardiac enzymes with normal coronary arteries on angiography.
The following points , as suggested by Myo Clinic are usually used as diagnostic criteria :

- Usually preceding stressful trigger is often present but not always present.
- Transient dyskinesia of the LV apical and /or mid segment.
- Regional wall motion abnormalities extending beyond a single epicardial vascular distribution.
- Absence of obstructive coronary disease or angiographic evidence of acute plaque rupture.
- New ECG changes (ST elevation / T-wave inversion) or moderate elevation in cardiac Troponin.
- Absence of pheochromocytoma and myocarditis.

6. Illustration by ECG



Source : : Prof. Dr. A.N. Rai , Former Prof. & Head Medicine and Prinicipal ANMMCH , Gaya, Bihar ; Chairman AIMS, Gaya

ECG of middle-aged non-diabetic female with h/o of hypertension and depression presenting as severe chest pain since 8 to 10 hours. Her Trop I was positive.
Coronary angiography showed normal coronary arteries.

ECG Findings :

- T-wave inversion in V2-V6 , more over V5-6 (consistent with more apical involvement) , extension of T-wave inversion also to limb leads I , aVL including II and III.
- A marked prolongation of QTc interval = 0.67 sec.

Discussion :
In the presence of such ECG changes with positive Trop I with normal coronaries in the background of depression as stressful event , there is a strong possibility of Takotsubo cardiomyopathy (Her follow up ECG did not show any changes at the time of discharge and report of cardiac echo not available).

7. Take-Home message

- ☐ Takotsubo cardiomyopathy is a special type of cardiomyopathy that is characterized by a transient stress induced reversible dysfunctioning with ballooning of a portion of the left ventricle of the heart , most often over its apex.
- ☐ The word 'Takotsubo' was coined from the name of a pot used by Japanese fishermen to trap octopuses. The apical part of the heart changes its contour like a round pot with its narrow neck – very much similar to the octopus trapping pot
- ☐ **Pathophysiology**
    - Catecholamine surge ⟶ action on the apical sympathetic nervous system fibers ⟶ microvascular spasm with myocytes-dysfunctioning.
    - In 10%-50% cases of TCMP there is an underlying some degree of left ventricular outflow tract (LVOT) obstruction ⟶ Cardiogenic shock
- ☐ **Complications** are encountered in some patients , which may include the followings :
    - Mild to moderate Mitral regurgitation
    - Acute left heart failure
    - Dynamic left ventricular outflow tract obstruction, which may lead to CCF or cardiogenic shock.
    - LV mural thrombus
    - Ventricular arrhythmias
    - At times even rupture of the ventricular wall
- ☐ Takotsubo cardiomyopathy should be suspected in young men and women presenting with chest pain with the background of emotional or physical stress (However , about 90% of affected individuals are women , and 80% are women over the age of 50). ECG changes are usually with repolarizing abnormalities – ST and T changes (ST segment elevation lasts shorter whereas T-wave inversion lasts longer). It would be difficult to differentiate Takotsubo cardiomyopathy from acute coronary syndrome and suspected cases should be referred to hospital. Diagnosis can be confirmed by finding normal coronary arteries on coronary angiography.
  NB : The regional wall abnormalities appear first on the cardiac echo in most cases in changes in the ECG appear subsequently one day later
- ☐ The prognosis of Takotsubo cardiomyopathy is usually good , with a mortality rate of 0-8%. Most patients have uneventful recovery with regaining of normal ventricular function within 1-4 weeks and have a good long term prognosis.

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CARDIAC FOOTPRINTS : IDENTIFYING AVRD ON ECG

ECG

CARDIAC FOOTPRINTS : IDENTIFYING ARVD ON ECG

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OUTLINE

Introduction
Arrhythmogenic Right Ventricular Dysplasia (ARVD is a form of genetically induced cardiomyopathy , being characterized by progressive replacement of right ventricular myocardium with fibrofatty tissue.

Arrhythmogenic Substrate in ARVD

- ☐ A basic concept of desmosome
- ☐ Genetic mutations
- ☐ Destablization of cardiac myocytes with its pathological changes
- ☐ Changes at the molecular level (dysregulation of Ca^{++})

Electrocardiographic changes (including the concept of Fontaine leads)

ECG-related findings as a part of the diagnostic evaluation through 'Task Force criteria'

Take Home Message

References

Cardiac Footprints : Identiying ARVD on ECG

A Narrative Review

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The tapestry upon a cloth may appear artistic; yet, when its weaving begins to fray, it quietly narrates its own demise with a sigh—"Ah…"

The same holds true for Arrhythmogenic Right Ventricular Dysplasia (ARVD), where the cohesive fibres of the myocardium—predominantly within the right ventricle—gradually lose their unity and fall into discordance.

- ☐ **The hallmark structural change in ARVD is progressive fibrofatty replacement of the right ventricular (RV) myocardium, leading to thinning and regional dysfunction of the ventricular wall.**
- ☐ **The ECG, as a silent scribe, reflects this electrical instability through characteristic findings—most notably the epsilon wave, a small post-QRS deflection seen in the right precordial leads, signifying delayed activation of diseased myocardium.**

The arrhythmogenic propensity of ARVD may culminate in life-threatening ventricular arrhythmias and sudden cardiac death, thereby underscoring the critical need for timely recognition and precise diagnosis.

1. Introduction (Keypoints)

- Arrhythmogenic Right Ventricular Dysplasia (ARVD) is a form of genetically induced cardiomyopathy , being characterized by progressive replacement of right ventricular myocardium with fibrofatty tissue.
- At the molecular level , it involves a complex interplay directed at cellular remodelling of the concerned RV , which expresses itself by dysregulation of intracellular calcium (Ca++) handling mechanism.
- Calcium ions play a critical role in the contraction of cardiac myocytes and its dysregulated signalling in ARVD contributes to its arrhythmogenic behaviour.
- The combined impact of basically three elements over right ventricular myocardium is contributing to its very nomenclature as **'Arrhythmogenic Right Ventricular Dysplasia' (ARVD) :**
  - **Arrhythmogenic** , denoting its propensity to develop ventricular arrhythmia.
  - **Right ventricular** myocardium , being its main site of expression
  - **Dysplasia** here points towards fibrofatty infiltration of the right myocardium

  Hence , 'Arrhythmogenic Right Ventricular Dysplasia' compasses all these three elements to express itself as a separate cardiomyopathic entity.
- ARVD causes a cluster of characteristic ECG changes , they dictate more or less the electrical translation of its associated histopathological changes.

ARVD with its main characteristic changes on ECG : (brief review)

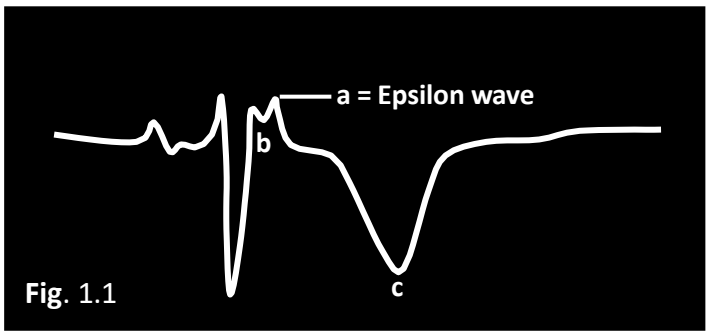


Fig. 1.1

Since architectural fibrofatty changes are mainly confined to RV , right precordial leads V1 through V3 record its associated characteristic changes on ECG :

a = Epsilon wave , a small deflection at the terminal portion of the QRS complex , looking like the shape of the Greek letter (ε) as a subtle wave.

b = Prolonged S wave upstroke

c = T-wave inversion over right precordial leads V1-V3

(The details would be discussed later on)

- ARVD causes symptoms being attributed to ventricular ectopic beats or sustained VT as a result of its arrhythmogenic behaviour. They typically present as palpitation , syncope or cardiac arrest precipitated by exercise.
  Notably to say , here the first presenting symptom may be even sudden cardiac death (there is usually a family history of sudden cardiac death in such cases)
  Over time, surviving patients develop features of right ventricular failure, which may progress to severe biventricular failure and dilated cardiomyopathy.

- The prevalence of ARVD is estimated to be 1 in 1000 to 1 in 5000 people. This disorder is usually remains undiagnosed because it can be difficult to detect in people with mild or no symptoms.

- Teenagers and young adults are more prone to ARVD compared to the old generation , though it is rare in both the groups. It can cause cardiac arrest (SCD) in young athletes.

- Risk involvement : The following patients are at high risk of sudden death if they have any one of the followings :
  - A history of syncope
  - Recurrent arrhythmias not suppressed by anti-arrhythmic drugs
  - A family history of cardiac arrest in first degree relatives

- Other terminology for ARVD :
  - Arrhythmogenic right ventricular cardiomyopathy (ARVC)
  - Right ventricular dysplasia

2. Arrhythmogenic substrate in ARVD

ARVD predisposes the sufferer to the paroxysmal episodes of ventricular arrhythmias and at times sudden cardiac death. It becomes essential to understand its arrhythmogenic substrate in such cases. The resulting arrhythmogenic substrate in patient with clinical ARVD is complex having multiple interrelated factors in its background. The following points are essential to be kept in mind for the purpose :

- **Normally the cardiac myocytes are adherent to each other by the intercellular disc filaments – known as desmosome**. These desmosomes help in interconnecting cardiac cells with each other , specially when they undergo mechanical stress. Whenever desmosome lose its normal architech , the cells of myocardium get detached from one another and are liable to die. This happens particularly when the heart muscle is placed under stress (e.g. during vigorous exercise)

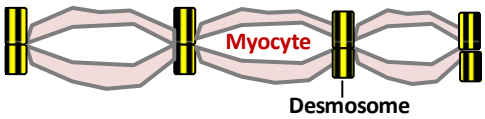


Fig. 1.2

Desmosomes attach cardiac myocytes to each other , providing strength to the myocardium as a whole and playing a pivotal role in sending electrical signaling between neighbouring cells.



Fig. 1.3

Whenever there is altered architect of desmosomes , usually induced by genetic mutations , the cardiac myocytes get detached from each other and are liable to die.

- **A number of genetic mutations adversely affect the structure and function of these desmosomes.** There have been reported mutations at least with 13 genes. Such genes are known as desmosomal genes because they provide signaling for making its components. By genetic mutations the desmosomal proteins affected include plakophilin, desmoplakin, and desmoglein. When the structure of these desmosomal protein are altered , the cardiac myocytes are prone to be damaged by mechanical stress (eg, from increased cardiac workload as from prolonged exertion).

  There are two patterns of inheritance:
  - Autosomal dominant: Where one parent has the mutation. Family members have a 50% chance of inheriting the abnormal gene.
  - Autosomal recessive: Where both parents have the mutation but no symptoms.

NB : ARVD may also be related to various non-genetic factors , e.g. viral or other inflammatory infections of the heart muscle or even other unknown causes. These factors can get operated independently or in conjunction with genetic predisposition.

☐ **Destabilization of cardiac myocytes brings a series of pathological changes :**

- Apoptosis (program cell death)
- Inflammation , enchanced fibrosis and loss of function of concerned myocytes
- Fatty replacement of myocardium
- Progressive dilatation and dysfunction of the concerned ventricle

The left ventricle is less commonly involved and the septum is relatively spared . and sometime LV involvement may preceed the RV involvement but with poor prognosis. Noteworthy to mention here that these changes usually begin in the epicardium and progress inward (the streched epicardium is the most stressed anatomical region)

☐ **Apart from the factors discussed so far there are certain changes also at the molecular level :**

- Calcium dysregulation with altered calcium handling leading to ventricular arrhythmias
  Abnormalities in calcium-handling proteins like the ryanodine receptor (**RyR2**) and sarcoplasmic reticulum Ca^{2+}-ATPase (**SERCA**) lead to intracellular calcium overload or leakage, which may lead to ventricular arrhythmias through the mechanism of 'delayed afterdepolarization' and reentry mechanism.

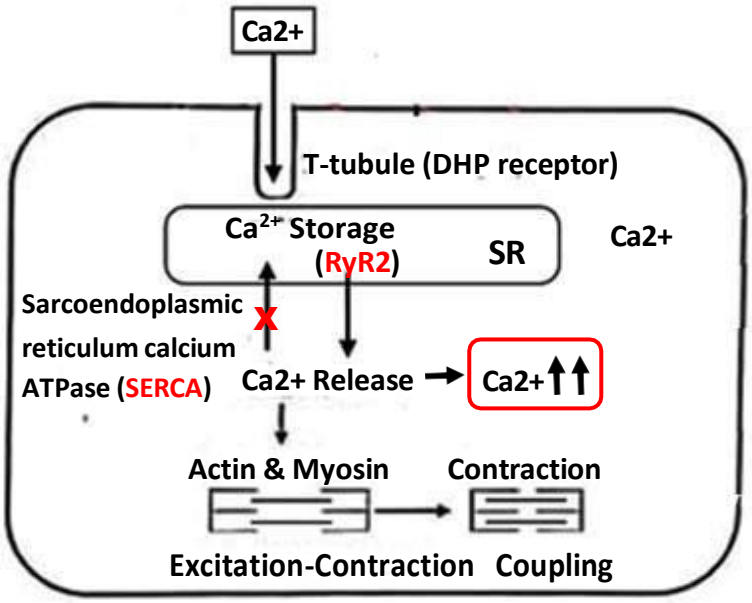


Fig. 1.4

This calcium malhandling may happen before the occurrence of gross structural changes in the myocardium , that's why , ventricular arrrhythmias may be the earlier manifestation.

- Dysregulated intracellular Ca2+ handling is particularly important during exercise, and a maladaptive response may explain the importance of exercise in the phenotypic expression and clinical outcomes in patients with ARVD.

Ref : This article is retrieved from the internet link given below :
Arrhythmogenic Right Ventricular Cardiomyopathy
Andrew D. Krahn MD [a], Arthur A.M. Wilde MD, PhD [b c], Hugh Calkins MD [d], Andre La Gerche MBBS, PhD [e], Julia Cadrin-Tourigny MD [f], Jason D. Roberts MD, MAS [g], Hui-Chen Han MBBS, PhD [a h]
April 2022
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3. Electrocardiographic changes

The typical ECG pattern of patients with arrhythmogenic right ventricular dysplasia (ARVD) resembles with that of incomplete RBBB with a negative T-wave over the right precordial leads V1 through V3 , with the additional terminal QRS deflection , known as Epsilon wave.

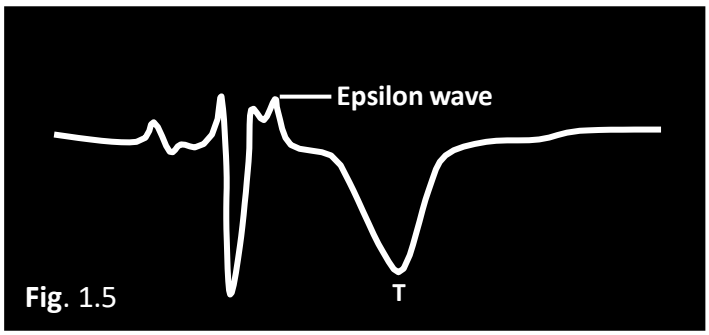


Fig. 1.5

It is not RBBB in true sense but appears so due to prolonged S-wave upstroke , as illustrated with the above sketch (a prototype wave pattern , observed over right precordial leads).

Basic of ECG changes :

- Fibrofatty changes in the right ventricular myocardium , delaying the conduction through the involved right ventricular myocardium
- The terminal late potetential (Epsilon wave) due to the late depolarization of the diseased myocardium
- Abnormal calcium handling and structural disorganization are central to the genesis of ventricular arrhythmias.

The following ECG changes have been observed with the cases of ARVD :

- ☐ T-wave inversion in right precordial leads V1-3 , in absence of RBBB (88% of patients) : this happens so due to the starting point of repolarization from the endocardium , in the opposite direction to that of exploring electrode → T-wave inversion.

- ☐ Epsilon wave (most specific finding) observed in 50% of cases. This Epsilon wave corresponds to a late potential due to delayed depolarization of diseased myocytes. It is probably a marker of advanced disease process.
- ☐ Localized QRS widening in V1-3 (>110 ms)
- ☐ Prolonged S-wave upstroke of ≥55 ms in V1-3. It is the most prevalent ECG criterion which correlates well with severity of the disease and induction of VT on ECG.
- ☐ Ventricular ectopy of LBBB morphology, with frequent PVCs > 1000 load per 24 hours
- ☐ Paroxysmal episodes of ventricular tachycardia (VT) with LBBB morphology.

NB : Some clinicians suggest recording of ECG rhythm strip lead II / V1 at double speed (50 mm / sec) and double amplitude (20 mV / mm) to have the detection of the ECG features with more clarity.

Fontaine leads : This would be worthwhile to mention here the significance of fontaine leads. They are specialized leads to increase the sensitivity of Epsilon wave detection.

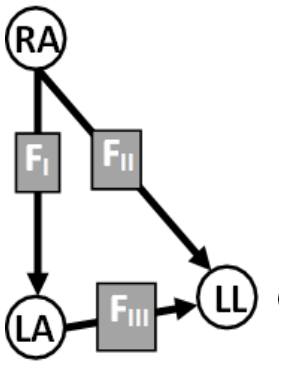


Fig. 1.6

Leads are placed as shown:

- Right Arm (**RA**) over the manubrium;
- Left Arm (**LA**) over the xiphoid process;
- and Left Leg (**LL**) in the standard V4 position (5th ICS MCL).

Instead of regular leads I, II, and III, there are now three *bipolar* chest leads that are termed FI, FII, and FIII. These record potentials developed in the right ventricle, from the RV infundibulum to the diaphragm (RV infundibulum is a funnl-shaped super portion of the right ventricle that acts as the outlet of right ventricular outflow tract – RVOT.

4. ECG-related findings as a part of the diagnostic evaluation through 'Task force criteria' for diagnosing ARVD

Here, only ECG-related criteria are mentioned for the purpose.

ECG-Related Major Criteria:

- Epsilon Wave in Leads V1-V3
- Ventricular Tachycardia (VT): Sustained or non-sustained VT of left bundle branch block (LBBB) morphology with a superior axis (negative QRS in leads II, III, and aVF, and positive in lead aVL). This pattern suggests origin from the inferior right ventricle.

ECG-Related Minor Criteria:

- Prolongation of the S-wave upstroke in leads V1-3
- T-Wave Inversions in lead V1-3
- Ventricular Tachycardia (VT): Sustained or non-sustained VT of LBBB morphology with an inferior axis (positive QRS in leads II, III, and aVF, and negative in lead aVL). This pattern suggests origin from the outflow tract of the right ventricle.

These ECG criteria are integral to the comprehensive assessment for ARVD, which also includes imaging studies, histological analysis, and genetic evaluation. The combination of these findings enhances the accuracy of ARVD diagnosis.

5. Take-Home Message

- ☐ Arrhythmogenic Right Ventricular Dysplasia (ARVD) is a form of genetically induced cardiomyopathy , being characterized by progressive replacement of right ventricular myocardium with fibrofatty tissue.
- ☐ Calcium ions play a critical role in the contraction of cardiac myocytes and its dysregulated signalling in ARVD contributes to its arrhythmogenic behaviour.
- ☐ ARVD causes symptoms being attributed to ventricular ectopic beats or sustained VT as a result of its arrhythmogenic behaviour. They typically present as palpitation , syncope or cardiac arrest precipitated by exercise.
- ☐ Teenagers and young adults are more prone to ARVD compared to the old generation , though it is rare in both the groups. It can cause cardiac arrest (SCD) in young athletes.
- ☐ Risk involvement : The following patients are at high risk of sudden death if they have any one of the followings :
    - A history of syncope
    - Recurrent arrhythmias not suppressed by anti-arrhythmic drugs
    - A family history of cardiac arrest in first degree relatives

- Abnormalities in calcium-handling proteins like the ryanodine receptor (**RyR2**) and sarcoplasmic reticulum Ca^{2+}-ATPase (**SERCA**) lead to intracellular calcium overload or leakage, which bring about a series of contraction-excitation coupling even leading to ventricular arrhythmias.

- ECG features :

  - T wave inversion in right precordial leads V1-3, in absence of RBBB (88% of patients)
  - Epsilon wave (most specific finding, seen in 50% of patients)
  - Localised QRS widening in V1-3 (> 110ms)
  - Prolonged S wave upstroke of 55ms in V1-3
  - Ventricular ectopy of LBBB morphology, with frequent PVCs > 1000 load per 24 hours
  - Paroxysmal episodes of ventricular tachycardia (VT) with LBBB morphology

- Fontaine leads are specialized leads to increase the sensitivity of Epsilon wave detection

- ECG related findings as a part of its diagnostic evolution through the Task Force Criteria for diagnosing ARVD should be taken into consideration.

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Personal Notes :