A Clinical And Electrocardiographic Arduous Diagnosis

Um diagnóstico clínico e eletrocardiográfico difícil

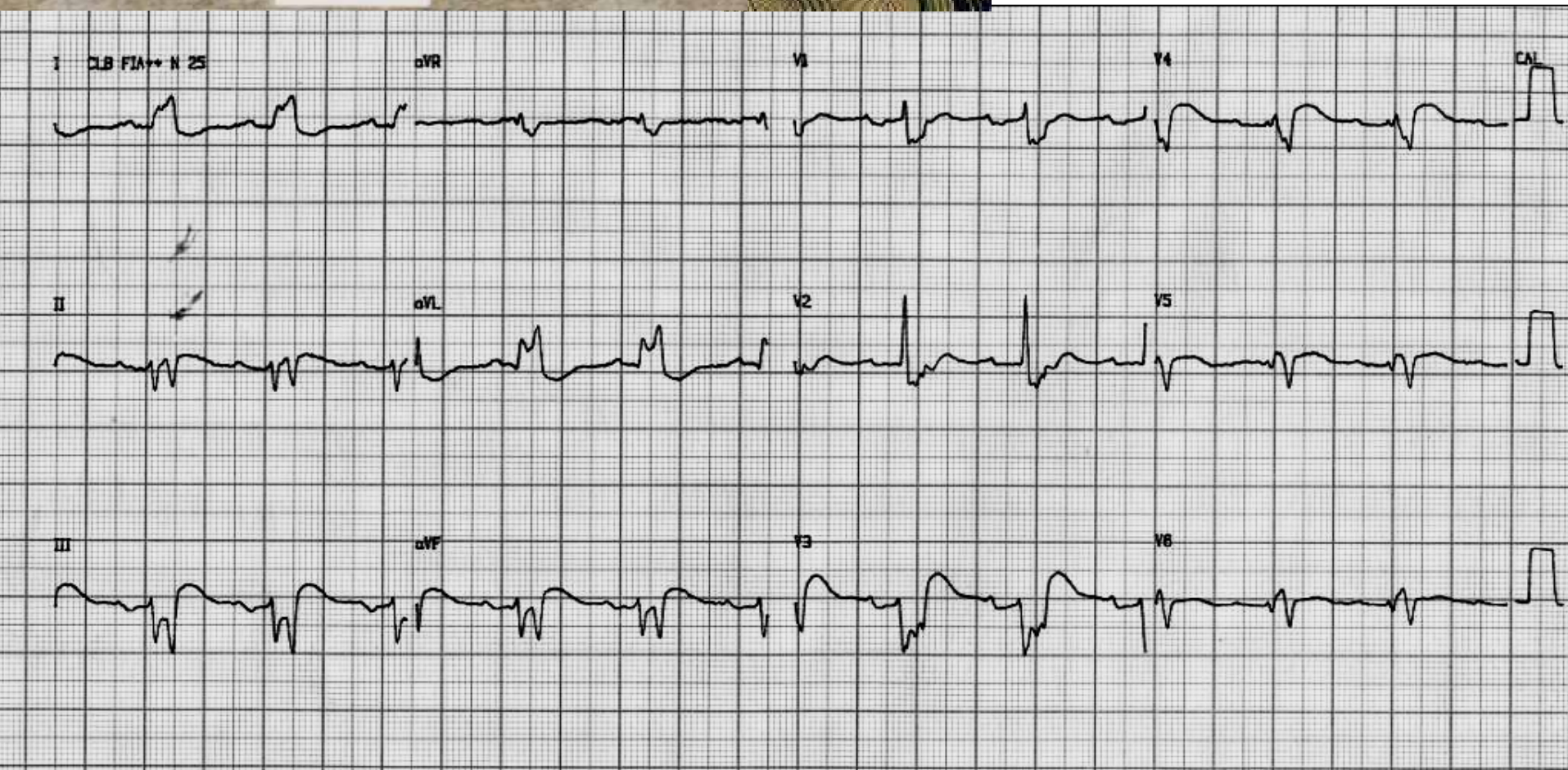
From Raimundo Barbosa-Barros " The Fox"

Final comments:
By Andrés Ricardo Pérez-Riera M.D. Ph.D.
"El Potro"

This is a 33-year-old woman admitted for evaluation for a heart transplantation. She was admitted with functional class IV of NYHA. There was an initial suspicion of muscular dystrophy due to neuromuscular alterations.
Regrettably, I don't have plenty of clinical data since the patient died and the family did not agree to an autopsy.
Transthoracic Echo; LV = 62/52; LA=53 mm, Mass 307 g, EF=31%, right chambers and pulmonary artery trunk without anomalies, the LV presented marked trabeculation in the lateral wall suggestive of Left Ventricular Noncompaction (LVNC) cardiomyopathy.
Mild mitral valve insufficiency.
Which is the ECG diagnosis?
Which is the clinical diagnosis: Muscular dystrophy? Noncompacted LV myocardium? Both?
Note: other family members died with similar symptoms (being young)
Raimundo **Barbosa-Barros M.D.**

Hola Maestro O que você acha deste ECG?
Infelizmente não tenho muitos dados clínicos pois o paciente veio a falecer e a família não aceitou a autopsia. Trata-se de uma mulher de 33 anos que foi internada para avaliação de transplante cardíaco. Admitida com ICC classe funcional IV.Havia uma suspeita inicial de distrofia muscular devido alterações neuro musculares.
ECO transtorácico; VE=62/52,AE=53mm,Massa 307g,FE=31%,câmaras direitas e TAP sem anormalidades,VE apresentando acentuada trabeculação na parede lateral sugestiva de miocárdio não compactado.I. mitral leve.
Distrofia muscular? Mioc. VE não compactado? Ambas?
Observação:outros membros da família faleceram com quadro semelhante(jovens)
Um abraço
Raimundo

The ECG diagnosis is not easy dear Watson.



Colleagues opinions

Spanish

Muy lindo trazado.Desde el punto de vista electrocardiográfico es como si la *masa ventricular estuviera partida en dos*. Un sector prácticamente normal y luego el sector anterior y lateral del ventrículo izquierdo con conducción lentísima y fraccionada.
La conducción dentro de las aurículas está también alterada.
El intervalo PR está alargado pero más a expensas de la duración de la onda P.Hacer diagnóstico de cardiomiopatian no-compactada en un solo sector del ventrículo izquierdo creo que es poco específico.
Es sabido que el trabeculado del mismo siempre es más fino que el del VD y cuando hay hipertrofia excéntrica y mas sectorial todo se parece. Yo me inclinaría más a pensar en una fibrosis idiopática. Hubiese estado bueno tener una resonancia.

Un abrazo: AC
Alejandro Cuesta M.D.Ph.D. Montevideo Uruguay

English
Very nice tracing. From the electrocardiographic point of view it is as if the ventricular mass is split in two. A virtually normal sector and then the anterior and lateral sector of the left ventricle with extremely slow and fractioned conduction.
Conduction within the atria is also altered.
The PR interval is wider but more at the expense of the P wave duration. I think making a diagnosis of noncompaction cardiomyopathy in a single sector of the LV is quite unspecific.
It is known that its trabeculation is always thinner than in the RV, and there is eccentric and more sectorial hypertrophy everything looks similar.
I would lean for idiopathic fibrosis. It would have been good to have a resonance.
Alejandro Cuesta M.D.Ph.D. Montevideo Uruguay

Dear Andrés
The ECG shows a very atypical LBBB +advanced Interatrial block + LA enlargement + left QRS axis deviation. The QRS suggests severe LV disease.
Best wishes
David H. Spodick, MD, FACC, MACP, FCCP, FAHA

Estimado Andrés: el ECG muestra un bloqueo de rama izquierda muy atípico + bloqueo interauricular avanzado + sobrecarga auricular izquierda+ desvio del eje del QRS para la izquierda. El QRS sugiere sever enfermedad del ventrículo izquierdo.

Having never seen a case of this disease I consulted Medline and Medscape, The anatomy, embryology and genetics are well explained, but ECG changes are described as "non-specific", This is why Dr Watson could not be of help. Anybody has other information about ECG?
Sem nunca ter visto um caso desta doença eu consultei e Medline Medscape. A anatomia, embriologia e genética são bem explicados, mas as alterações no ECG são descritos como "não específicas", É por isso que o Dr. Watson não poderia ser de ajuda. Alguém tem outras informações sobre ECG?
Como nunca había visto un caso de esta enfermedad he consultado Medline y Medscape, la anatomía, la embriología y la genética están bien explicadas, pero cambios en el ECG se describen como "no específica", es por eso el doctor Watson no podía ser de ayuda. ¿Alguien tiene otra información sobre el ECG?

Borys Surawicz M.D.
Professor Emeritus
Indiana University School of Medicine
Indianopolis, Indiana

Desde el punto electrocardiografico muestra uns depolarization del ventriculo izquierdo muy lenta durando 120 ms depolarizarse , en DI dando la impresion de ser unBRI,pero no lo es BRI Con este electro el diagnóstico es de dilatacion vent izquierda idiopatica.

Eventualmente podria ser una cardiomiopatias post partum (si tuvo hijos), com desenlace dramatico en poco tiempo En adultos podria pensarse una cardiopatia isquemica ,pero en esta mujer joven es dificil. Unicamente que fuese una anomalia coronaria muy complicada.Tambien podria ser una miocardiopatia por mutacion de algunas de las proteinas del sarcomero (una biopsia del ventrículo izquierdo podria darnos una respuesta) y en especial que parece que tiene concomitantemente una miopatia periférica

Las cardiopatias infiltrativas se puede descartar , ya que estas generalmente se dilatan despues de una hipertrofia del vent izquierdo y evolucionan en patchs (me olvide como se dice en espaniol)

El ECG muestra una afectacion general del musculo cardiaco, con conduccion intraventricular muy lenta, y complejos con voltaje muy disminuidos, y unicamente el area del septo superior, pudo remodelada,,en forma no eficiente ,con la cara lateral.

Un caso dramatico una pena no tener mas datos

Un fraternal abrazo

Samuel Sclarovsky

From the electrocardiographic point of view, the patient shows very slow LV depolarization lasting 120 ms, in DI giving the impression of being LBBB, but it is not. With this ECG, the diagnosis is of idiopathic LV dilatation.

It could be post-partum cardiomyopathy (if she had children) with a dramatic outcome in a very short time.

In adults, we could think ischemic heart disease, but in this young woman it is difficult. Only if it was a very complicated coronary artery anomaly. It could also be cardiomyopathy by mutation of some of the sarcomere proteins (a biopsy of the LV could yield an answer) and especially it seems that there is concomitant peripheral myopathy.

Infiltrative heart diseases can be ruled out, since these generally dilate after LV hypertrophy and evolve in patches.
The ECG shows general involvement of the cardiac muscle with very slow intraventricular conduction and complexes with very decreased voltages, and only the area of the upper septum could remodel in a non-efficient way with the lateral side.
A dramatic case; too bad we don't have any more data.
Warm regards,
Samuel Sclarovsky Israel

Querido Samuel apenas para recordarle Hay 2 datos relevantes no mencionados en sus comentários:
Que el ventriculo tenia traveculaciones sugestivas de corazón no compactado
Que havia sospecha clinica de enfermedad neuro-muscular concomitante.
Andres.

Dear Samuel,
I would just like to remind you there are 2 relevant data not mentioned in your comments:
That the ventricle had trabeculations suggestive of noncompaction heart That there was the clinical suspicion of concomitant neuro-muscular disease.
Andrés

Dear friend, Prof. Andrés Ricardo Pérez Riera, You are right, according to the echo report on noncompaction cardiomyopathy this should be considered. There is another less rare heart disease, but it also displays signs of hypertrophy: eosinophilic ones.
In my analysis I considered the possibility of a mutation of the sarcomere proteins, due to the presence of a peripheral myopathy.
Warm regards,
Samuel Sclarosky

Querido amigo Profesor Andrés Ricardo Pérez Riera usted tiene razon, segun el informe del ecocardiograma sobre non compacting cardiomyopathyy deberia traerlo a consideracion.
Hay otra cardiopatia menos rara , pero viene tambien con signos hipertrofia son las eosinofilicas
En mi análisis puse en consideracion la posibilidad de una mutación de las proteina del sacomero , por la presência de una miopatia periférica
un fraternal abrazo
Samuel Sclarovsky

Caros “El Potro” e “The Fox” e demais amigos do Fórum A maioria dos raros relatos de casos (algumas centenas) de miocardio não compactado o descrevem como apical, associado ou não a atresia pulmonar ou obstrução da via de saída do VD e anomalia na origem da CE (Elias e cols.Miocardio não compactado isolado -Arq Bras Cardiol volume 74, (nº 3), 2000). A localização lateral do eco não afasta miocárdio não compactado, mas pergunto se foi realizado imunologia p/ Chagas, cuja miocardiopatia não é trabecular e tambem não lateral e sim apical ou póstero-inferior. Já há relato de caso da associação. O ECG chama atenção pela fragmentação do QRS e pela onda R alta em V2.

O Vectocardiograma, se realizado, teria substancial contribuição no caso, não?
Abraços
Adail

Dear “El Potro” and “The Fox”, and the rest of the friends of the Forum, Most of the rare narrations of the cases (several hundreds) of noncompaction myocardium, describe it as apical associated or not to pulmonary atresia or RVOT obstruction and anomaly with an origin in the left coronary artery (Elias e cols.Miocardio não compactado isolado -Arq Bras Cardiol volume 74, (nº 3), 2000). The lateral location of the echo does not rule out noncompaction myocardium, but I wonder if immunology for Chagas disease was made, the cardiomyopathy of which is not trabecular and is also not lateral but apical or postero-inferior. There is already a case of association. The ECG is remarkable for the QRS fragmentation and by the high R wave in V2.
The vectorcardiogram, if performed, would be a substantial contribution, right?
Warm regards,
Adail Paixão Almeida M.D. Vitória da Conquista Bahia Brazil

Estimado Raimundo como siempre sus casos y especialmente este ECG es espectacular. Coincido en lo que plantea Samuel, y como todos saben las enfermedades neuromusculares presentan o se asocian con afectación cardiaca en forma significativa, siendo el espectro amplio pero preferentemente el sistema de conducción, pudiendo en algunos casos ser esta la principal causa de muerte súbita por BAV completo. La asociación entre enfermedades neuromusculares y trabeculación del VI (no compactación) también se encuentra descripta, en casos aislados, pero la asociación existe sin dudas. Creo que particularmente este ECG es para mostrarlo en algún ítem de ECG interesting, he revisado la literatura y hemos escrito algunas cosas sobre el tema y yo al menos es la primera vez que veo como imagen este hermoso ECG. Raimundo se sabe que enfermedad neuromuscular presentaba la paciente y sus familiares?Saludos y felicitaciones, si quiere Raimundo metemos manos a la obra, como siempre.
Pancho

English
Dear Raimundo, as usual your cases, and especially this ECG, are spectacular.I agree with what Samuel proposed, and as you all know neuro-muscular diseases present or are associated to cardiac affections in a significant way, with a wide spectrum but preferably the conduction system. In some cases this could be the main cause of sudden death by complete AV block. The association between neuro-muscular diseases and LV trabeculation(noncompaction) is also described in isolated cases, but the association exists undoubtedly. I think that this ECG in particular is to show some interesting ECG item. I have reviewed literature and we have written some stuff about this issue, and for me this is at least the first time I see this beautiful ECG as an image. Raimundo, do you know that neuro-muscular disease did the patient and her relatives present?

Francisco Femenía M.D.
Arrhythmia Unit. Cardiology Department. Hospital Espanol de Mendoza. Argentina.

Algunas referencias: Some references

1. Acquired left ventricular hypertrabeculation/noncompaction in myotonic dystrophy type 1. Finstere J et al. Int J Cardiol 2009
2. Neuromuscular implications in left ventricular hypertrabeculation noncompaction. Finstere J et al. Int J Cardiol 2006
3. Left ventricular hypertrabeculation noncompaction with and without neuromuscular disorders. Stollberger C et al. Int J Cardiol 2004
4. Slowly progressive conduction system disturbance in a patient with polymyositis. Femenía F et al. Anadolu Kardiyol Derg 2011
5. Progressive conduction disturbance in myotonic dystrophy. Palazzolo J et al. Cardiol J 2011

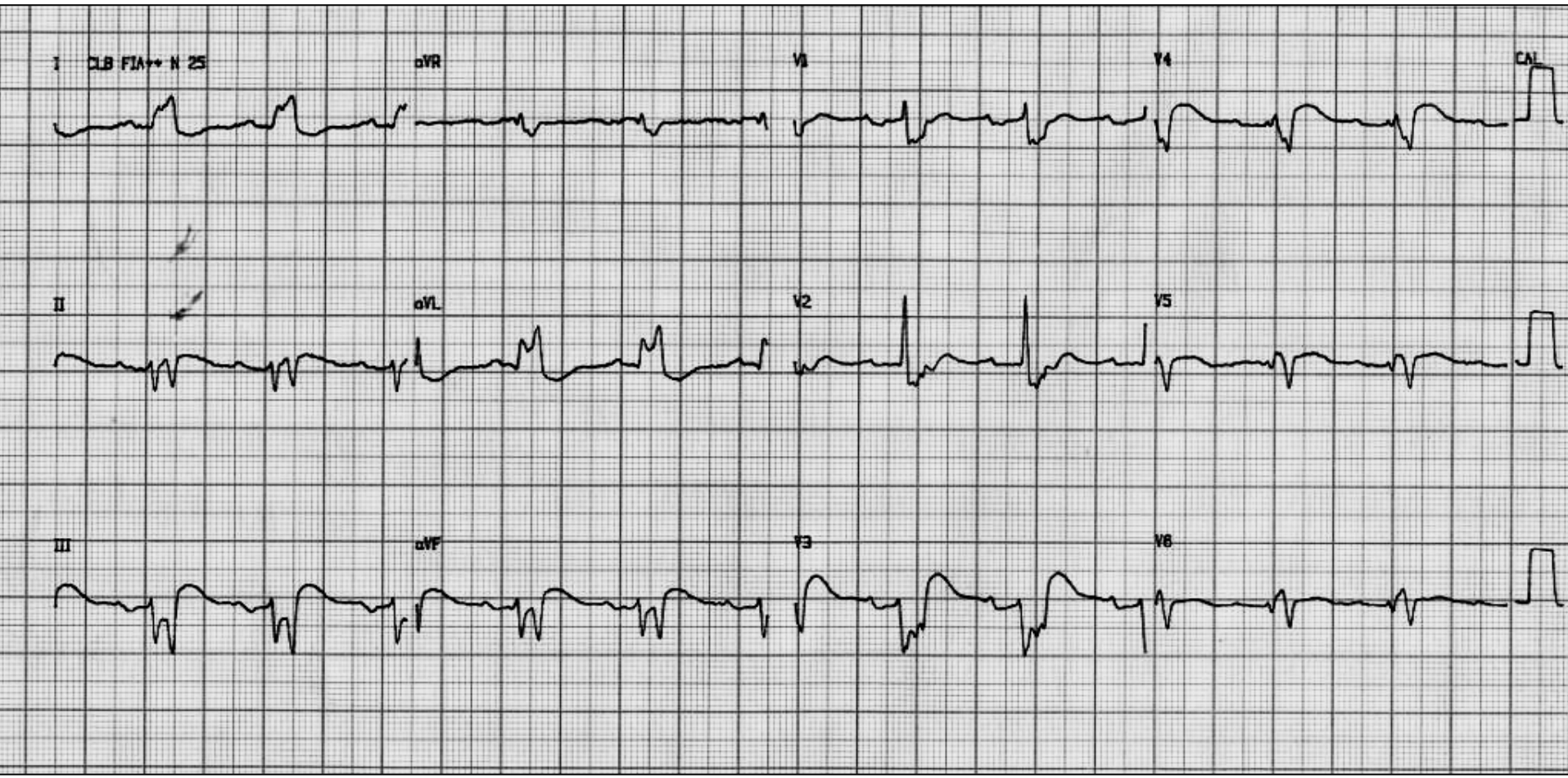
de Campos para o Prof Andrés.
Bom dia !Grande abraço.

Trata-se de ritmo sinusal com bloqueio AV de primeiro grau, com significante distúrbio de condução intraventricular que acomete ambos os ventrículos (parecendo miocardiopatia não impactada , repercutindo na não amplitude vetorial clássica. Sem vetor ,sem definição de agrupamento muscular ,sem FE) com QRS de 180 ms, com predominante BCRE ,com alteração severa na repolarização ventricular, Sugere grande destruição de massa muscular,com múltiplas áreas inativas.
Como estão os eletrólitos? É importante
ECG de qualquer idade desta paciente? ou,de familiar que faleceu jovem.
Há um padrão ECG genômico da família?
Falaremos,
Lourival
lourivalcampos@yahoo.com.br

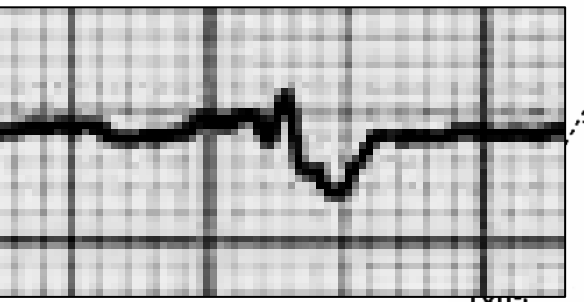
This is sinus rhythm with first degree AV block, with significant intraventricular conduction disorder that affects both ventricles (like Left Ventricular Noncompaction (LVNC) cardiomyopathy, resulting in non-classical amplitude vector. Without vector, with no definition of muscle group without EF) with QRS duration of 180 ms, predominantly LBBB with severe changes in ventricular repolarization, suggested massive destruction of muscle mass with multiple inactive areas.
How are electrolytes? It is important
ECG of any patient of this age? or a family member who died young.
There is a genomic pattern family?
We'll talk,.....
Lourival Campos M.D. Sao Paulo Brazil

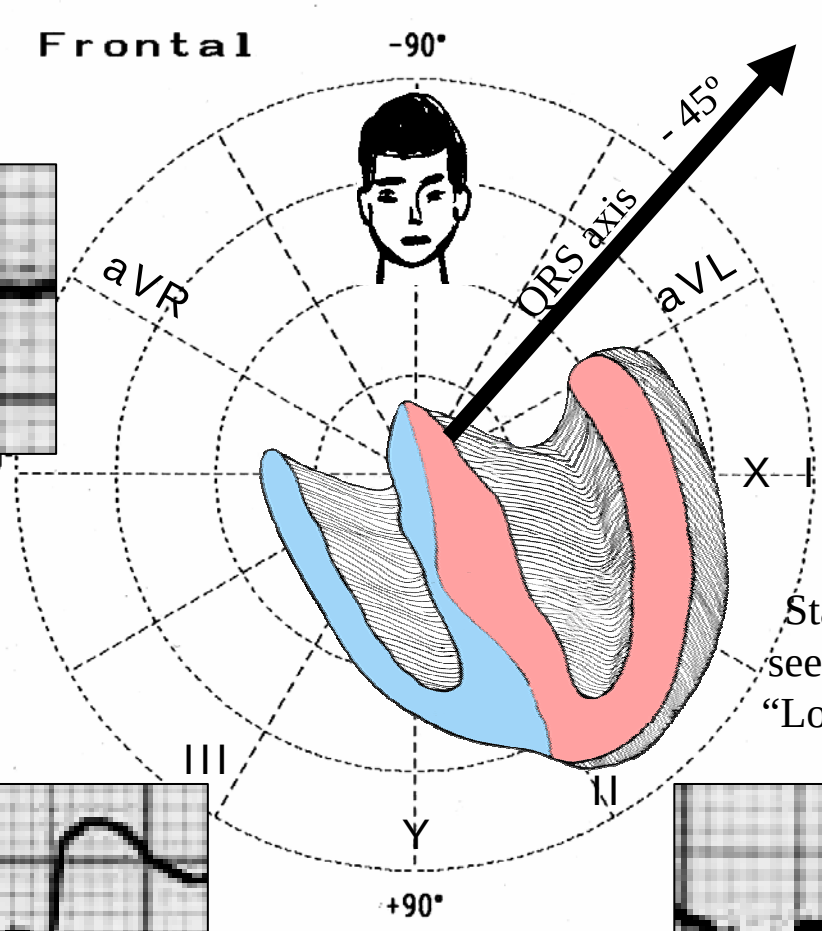
Final comments

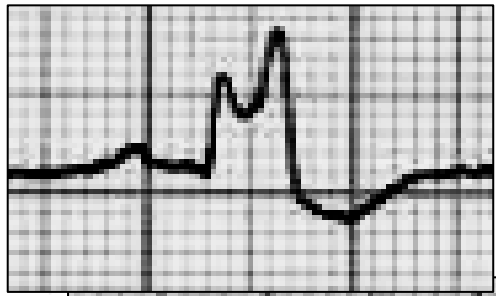
By Andrés Ricardo Pérez-Riera M;D.Ph.D.

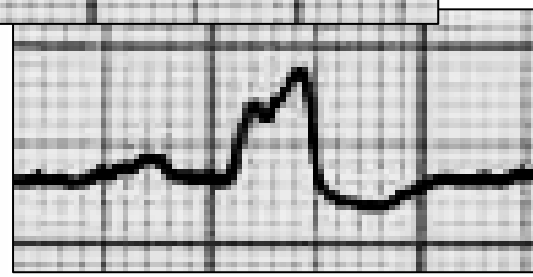

I CLB FIA++ N 25
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6

Extreme left axis QRS deviation



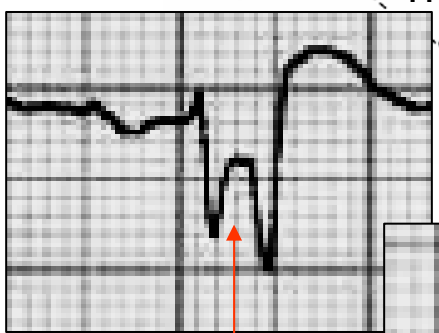


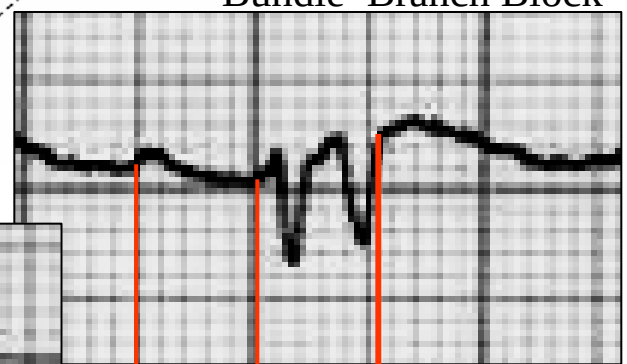


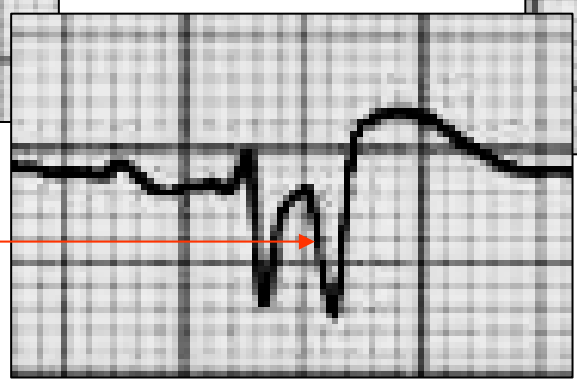


Standard I and aVL QRS patterns seems very broad Complete LBBB "Look-like LBBB" or resemble Left Bundle–Branch Block

P axis + 25º







Wide and fragmented QRS complexes (f-w-QRS)
See comments in next slide.

QRSd= 205ms: very broad

PR-interval = 200ms

Fragmented QRS complexes (fQRS) were proven to be associated with the prognosis of several heart diseases. However, no data is available regarding fQRS in Left Ventricular Non-Compaction (LVNC) cardiomyopathy, in which the outcome varies greatly and a simple yet practicable prognostic predictor is needed. With the purpose of to determine the prognostic value of fQRS in LVNC patients Ning et al(1) studied 64 LVNC patients.

Fragmented narrow QRS (f-QRS) included single or multiple notches in the R or S wave in at least 2 contiguous ECG leads and QRS duration < 120 ms.

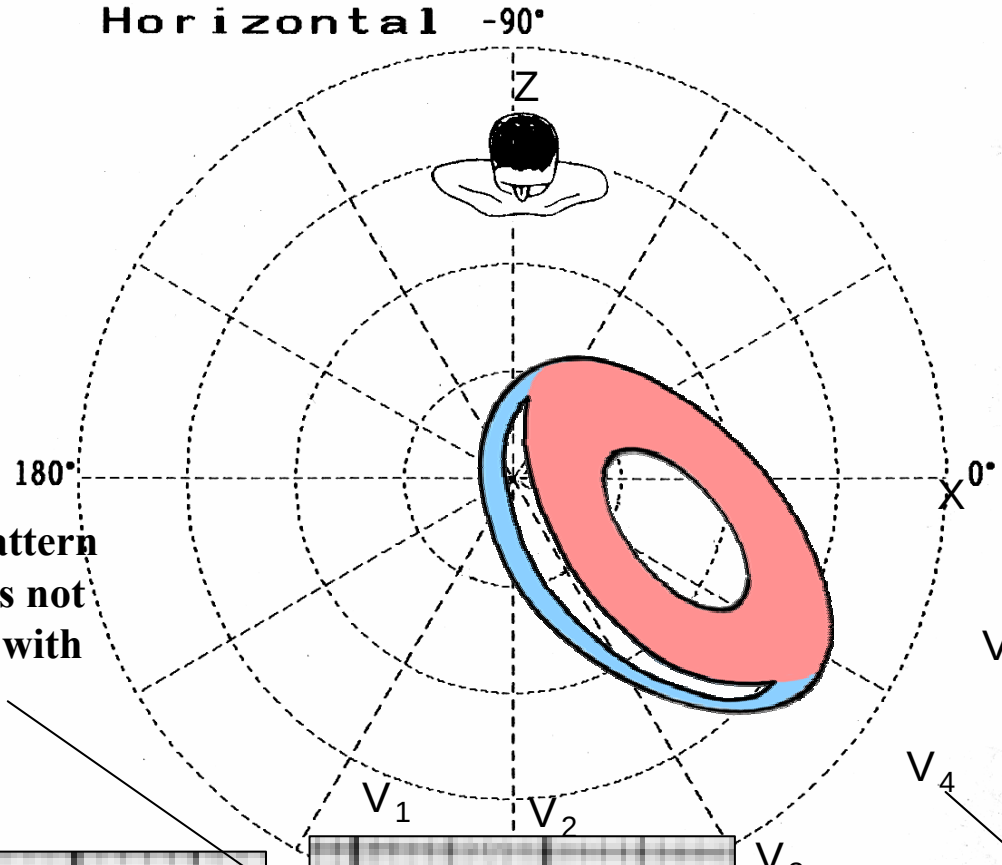
Fragmented wide QRS (f-wQRS) included more than 2 notches and QRS duration ≥120 ms.

RESULTS:
f-QRS and f-wQRS was present in 24 (38%) and 7 (11%) patients respectively. During follow-up, 13 patients died and 7 patients underwent heart transplantation.
Kaplan-Meier analysis revealed that compared with the non-f-nQRS group, the f-QRS group was associated with a significantly lower survival (P = 0.005).
The f-wQRS group also demonstrated a substantially lower survival as compared with the non-f-wQRS group (P = 0.02).
Multivariate analysis indicated f-QRS was an independent predictor of all-cause mortality.

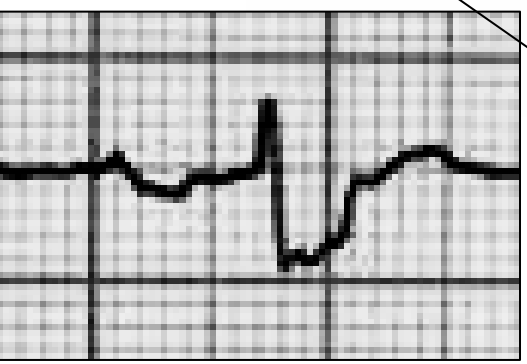
The authors concluded that in Left Ventricular Non-Compaction patients, the presence of f-QRS has significant prognostic value and may provide a valid method of risk stratification.

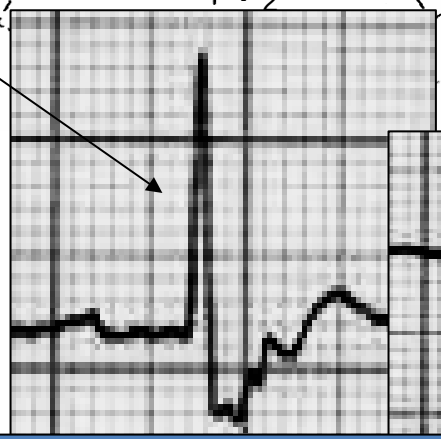
1. **Ning XH, Tang M, Chen KP, Hua W, Chen RH, Sha J, Liu ZM, Zhang S. The Prognostic Significance of Fragmented QRS in Patients With Left Ventricular Noncompaction Cardiomyopathy. Can J Cardiol 2012 Jul;28:508-514.**

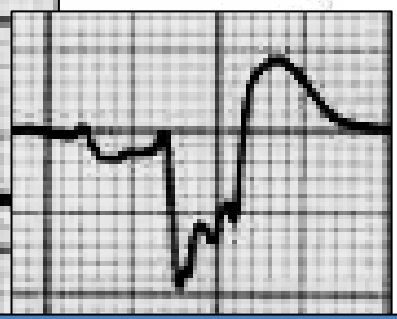
Nonspecific intraventricular conduction disturbance or Nonspecific intraventricular conduction delay



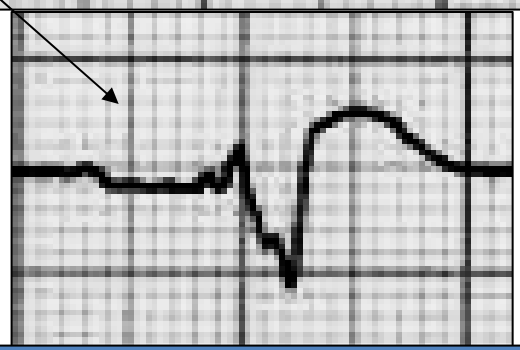
This QRS pattern (Rs) in V_2 is not compatible with LBBB



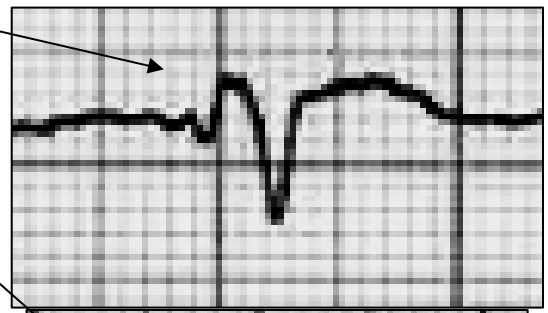




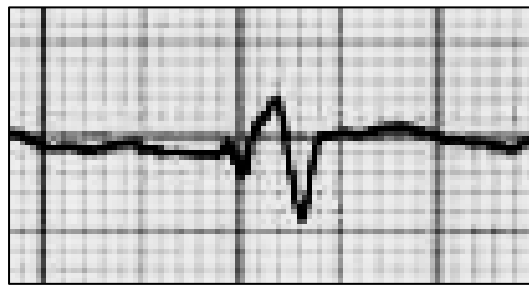
V_4



V_5



V_6



1. Mirvis, DM, Goldberger, AL. Electrocardiography. In: Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine, 9th ed, Bonow, RO, Mann, DL, Zipes, DP, Libby P (Eds), W.B. Saunders Company, Philadelphia 2011.

Nonspecific intraventricular conduction disturbance or Nonspecific intraventricular conduction delay

Definition: Atypical QRS widening ≥110ms that does not fit the ECG criteria either LBBB, RBBB or Wolff-Parkinson-White syndrome caused by complex delays mainly in the LV parietal wall, mural, myocardium or Purkinje arborization, eventually associated with stem or divisional block or a combination of previous ones.

Synonymous:

1. **Nonspecific intraventricular block or Nonspecific IV block**
2. **Unspecified intraventricular block or Unspecified IV block**
3. **Intraventricular conduction delay or IV conduction delay**
4. **Arborization block**
5. **Peri-infarction block**
6. **Mural blocks**
7. **Parietal blocks**

Possible etiologies

1. **Severe hyperkalemia**
2. **Drugs that block sodium channels, such as Class IA (quindine), IB and IC Propafenone**
3. **Tricycles antidepressant**
4. **Infiltrative granulomatous: amiloidosis, sarcoidosis**
5. **Chagas myocarditis**
6. **Myocarditis**
7. **Cardiomyopathies example: Left Ventricular Non-Compaction (LVNC) cardiomyopathy. Non-compaction cardiomyopathy (NCC),**
8. **Intra-infarction, intramural.**
9. **Surgical right ventriculotomy**

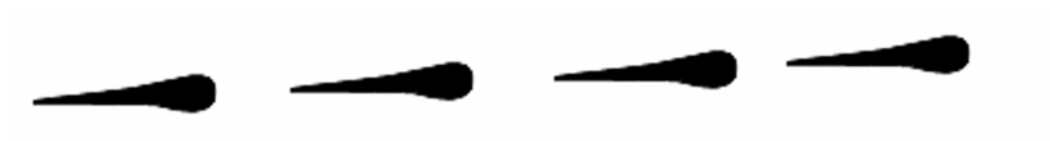
Meaning According To The Location Of The Conduction Delay

INITIAL QRS CONDUCTION DELAY = *Pre-excitation, WPW syndrome/ delta wave.*
END and MIDDLE CONDUCTION DELAY = **Stem** *Complete Left Bundle Branch Block.*
END CONDUCTION DELAY = **Predivisional** *Complete Right Bundle Branch Block or IRBBB.*
UNIFORM CONDUCTION DELAY = *Hyperkalemia; quinidine or other drugs that block sodium channels, tricycles antidepressant drugs; intra-infarction, intramural or non-specific* intraventricular conduction disturbance or nonspecific intraventricular conduction delay.

Determination of conduction velocity of stimulus

In vectorcardiography, the greater or the lesser distance between dashes indicates the greater or the lesser conduction velocity in the area. Thus, when they are very close to each other, it indicates the presence of conduction delay. To consider the phenomenon as true, it is necessary for it to be evident in at least 2 planes.

Separate dashes = more dromotropism Larger speed. Faster conduction



Very close dashes = less dromotropism.Smaller speed. Slower conduction

Location of delay in right or left bundle branch block, Wolff-Parkinson-White, and the causes of uniform intraventricular conduction delays. Knowledge of conduction velocity by the distance between dashes in VCG.

TOPOGRAPHIC TYPES OF LEFT BUNDLE BRANCH BLOCK (LBBB)

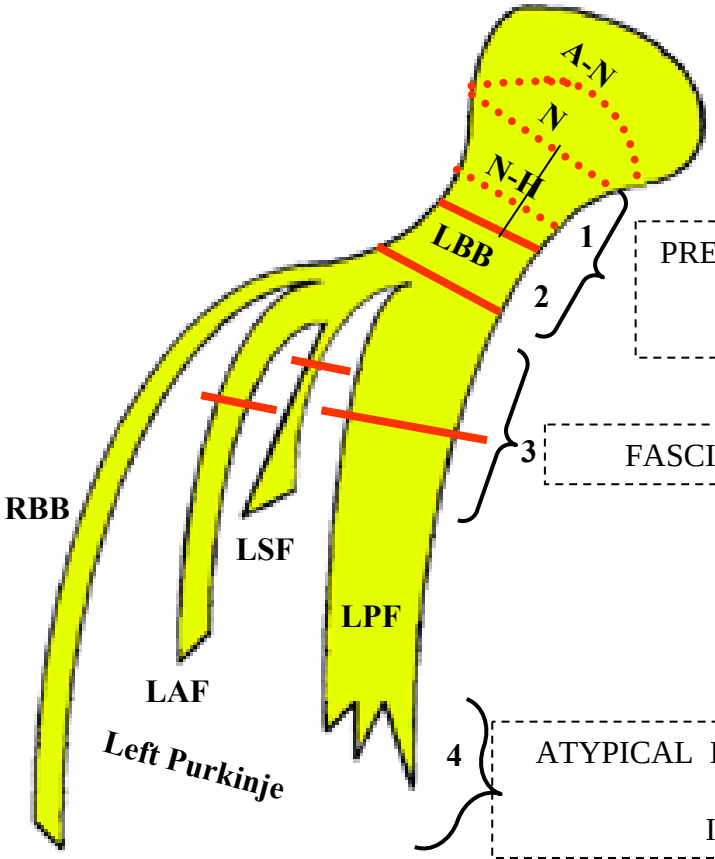
CONCEPT: LBBB, left His system global block or left ventricle global block, is any delay in left ventricle (LV) activation as a consequence of a dromotropic disorder located in one or more of the following sites:

1. **Left His bundle**
2. **Stem (Truncus) of Left Bundle Block (LBBB):** 1 and 2 are known as pre-divisional or membranous LBBB.
3. **Fascicular or divisional LBBB of the His bundle concomitantly:** left anterior fasciclicular (LAFB), left posterior fascicular (LPFB), and left septal or middle-septal fascicular block (LSFB); This type is known as fascicular or divisional LBBB.

4. **LEFT PURKINJE GLOBALLY APPROACHED**: This is known as parietal, Purkinje, intramural or intramyocardial pseudo LBBB or atypical LBBB. This is not a truly LBBB. It is considered a Nonspecific intraventricular conduction disturbance or Nonspecific intraventricular conduction delay It is caused by complex delays mainly in the LV parietal wall, mural, myocardium or Purkinje arborization, eventually associated with stem or divisional block or a combination of previous ones.

Concept of left bundle branch block and its proximal or predivisional, divisional or fascicular and Purkinje-muscular or intramyocardial location.

1) **Left portion of His bundle**
2) **Stem of Left bundle branch**
3) **Divisions or fascicles of LBB.**
4) **Parietal, intramural, Purkinje or intramyocardial block It is considered a Nonspecific intraventricular conduction disturbance or Nonspecific intraventricular conduction delay**

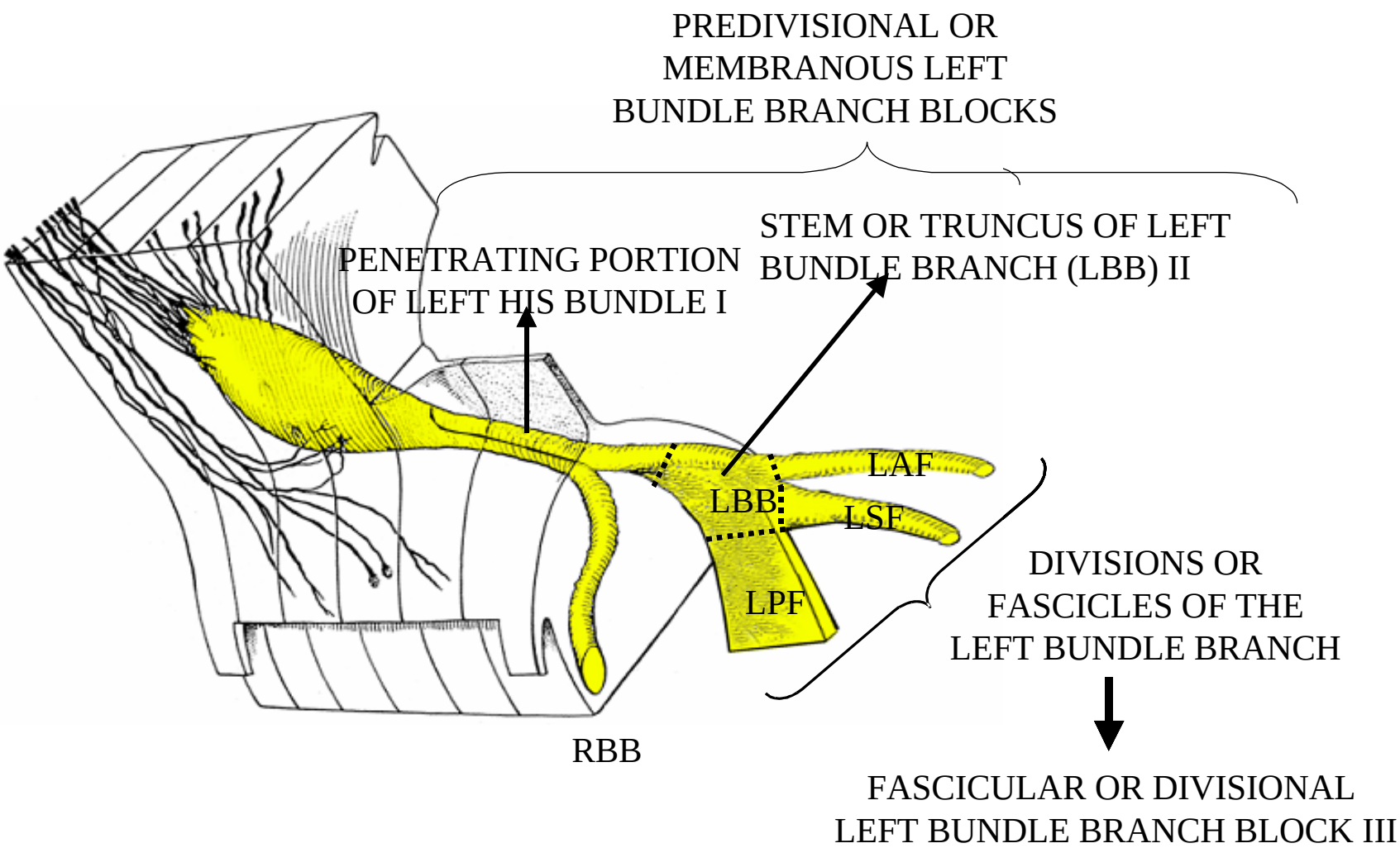


A-V N: Atrio-Ventricular Node.
A-N: Atrio-Nodal region of the A-V node.
N: Nodal region of A-V Node.
N-H: Node-His region of the A-V Node.
RBB: Right Bundle Branch.

LBB: Left Bundle Branch.
LAF: Left Anterior Fascicle.
LSF: Left Septal Fascicle.
LPF: Left Posterior Fascicle.

Outline of the three portions in the left bundle branch block and nomenclature of the intraventricular His system.

CLBBB ACCORDING TO TOPOGRAPHY



Outline that shows the left bundle branch block according to topography.

Clinical-therapeutic significance of parietal, Purkinje, intramural, intramyocardial or pseudo LBBB or atypical LBBB.

Cardiac Resynchronization Therapy (CRT) is effective in reducing clinical events in systolic heart failure patients with a wide QRS. Retrospective studies suggest only patients with QRS prolongation due to a LBBB benefit from CRT. Sipahi et al (1) with the objective to examine this by performing a meta-analysis of all randomized controlled trials of CRT. While CRT was very effective in reducing clinical events in patients with LBBB, it did not reduce such events in patients with wide QRS due to other conduction abnormalities such as Parietal, intramural, Purkinje or intramyocardial block It is considered a Nonspecific intraventricular conduction disturbance or Nonspecific intraventricular conduction delay.

Normal ventricular depolarization occurs after an impulse traverses the atrioventricular (AV) node and the bundle of His. This specialized conducting tissue splits into two main branches, the right and the left bundle branches, that rapidly transmit depolarization wavefronts to the right and left ventricular myocardium, respectively, via the Purkinje fibers.

The main stem left bundle branch trifurcates into three primary subdivisions:

A left anterior fascicle (LAF);

A left septal or left median (middle or septal) fascicle (LSF) and

A left posterior fascicle (LPF).

The depolarization wavefronts spread through the ventricular wall, from endocardium (inner layer), mid-miocardium, to epicardium (outer layer), triggering intracellular calcium release and myofilament contraction (electromechanical coupling).

1. **Sipahi I, Chou JC, Hyden M, Rowland DY, Simon DI, Fang JC.Effect of QRS morphology on clinical event reduction with cardiac resynchronization therapy: meta-analysis of randomized controlled trials. Am Heart J. 2012 Feb;163:260-7.e3.**

Cardiac resynchronization therapy (CRT) has emerged as an attractive intervention to improve left ventricular mechanical function by changing the sequence of electrical activation. Unfortunately, many patients receiving CRT do not benefit but are subjected to device complications and costs. Thus, there is a need for better selection criteria. Current criteria for CRT eligibility include a QRS duration >120 ms. However, QRS morphology is not considered, although it can indicate the cause of delayed conduction.

Studies have suggested that only patients with LBBB benefit from CRT, and not patients with **right bundle branch block or nonspecific intraventricular conduction delay**.

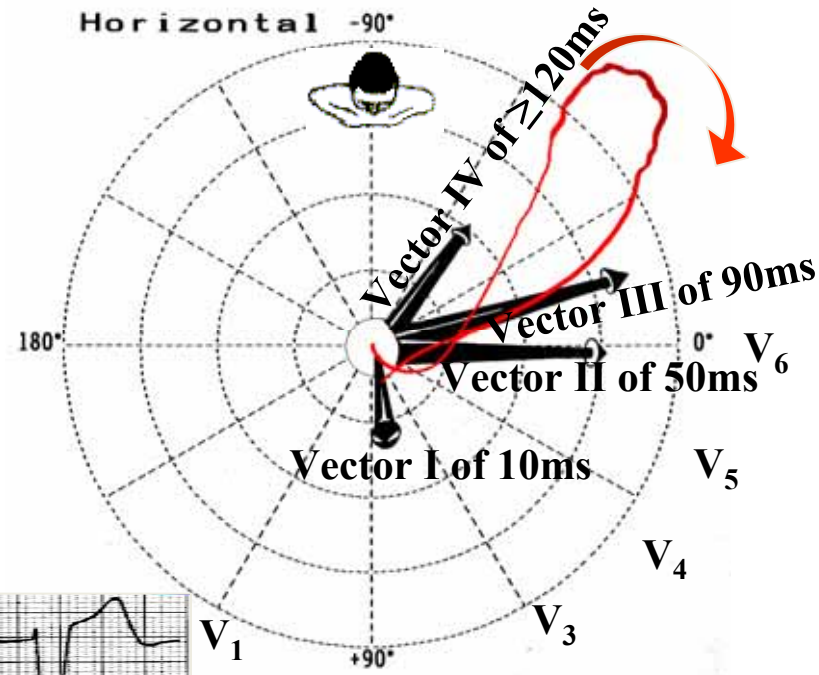
Three key studies over the past 65 years have suggested that 1/3 of patients diagnosed with LBBB by conventional ECG criteria may not have true complete LBBB, but likely have a combination of LVH and LAFB. On the basis of additional insights from computer simulations, the investigators propose stricter criteria for complete LBBB that include: A **QRS duration >140 ms for men and >130 ms for women**, along with mid-QRS notching or slurring in >2 contiguous leads.

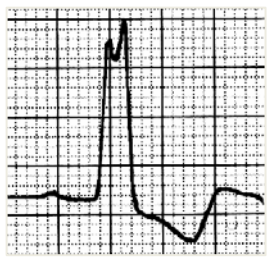
1. **Surawicz B, Childers R, Deal BJ, Gettes LS, Bailey JJ, Gorgels A, Hancock EW, Josephson M, Kligfield P, Kors JA, Macfarlane P, Mason JW, Mirvis DM, Okin P, Pahlm O, Rautaharju PM, van Herpen G, Wagner GS, Wellens H, American Heart Association Electrocardiography and Arrhythmias Committee, Council on Clinical Cardiology, American College of Cardiology Foundation, Heart Rhythm SocietySOAHA/ACCF/HRS recommendations for the standardization and interpretation of the electrocardiogram: part III: intraventricular conduction disturbances: a scientific statement from the American Heart Association Electrocardiography and Arrhythmias Committee, Council on Clinical Cardiology; the American College of Cardiology Foundation; and the Heart Rhythm Society. Endorsed by the International Society for Computerized Electrocardiology. J Am Coll Cardiol. 2009;53(11):976.**
2. **Gettes LS, Kligfield P. Should electrocardiogram criteria for the diagnosis of left bundle-branch block be revised?J Electrocardiol. 2012 Jul 16. [Epub ahead of print]**

ECG/VCG CORRELATION IN HORIZONTAL PLANE, IN STEM LEFT BUNDLE BRANCH BLOCK

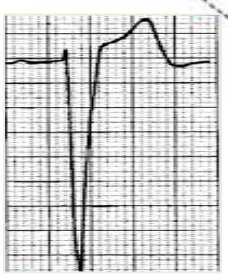
Outline that shows the direction and magnitude of the four vectors that represent the ventricular depolarization in the Horizontal Plane and the QRS morphologies in V_1 and V_6

Characteristically the QRS loop is inscribed Clockwise in HP

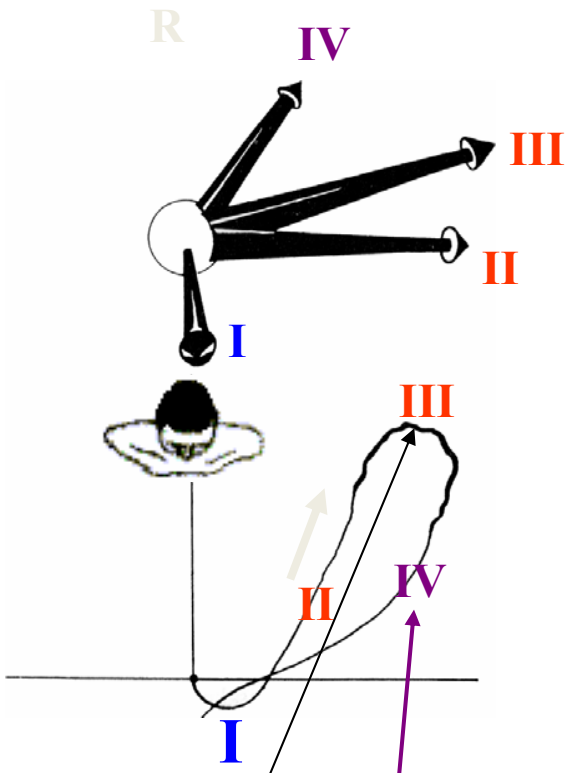




R pattern



rS or QS pattern



Vectors III and IV with middle and end conduction delay

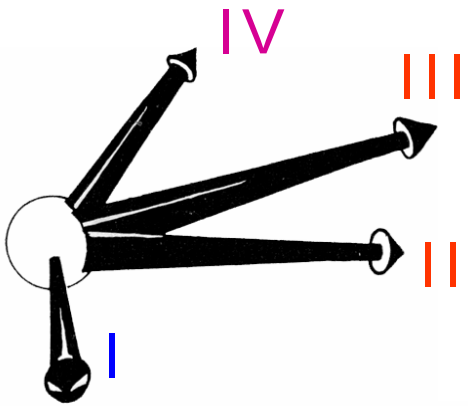
THERE ARE THREE MAYOR COMPONENT IN VENTRICULAR ATIVATION PROCESSES IN UNCOMPLICATED TRULLY LBBB:

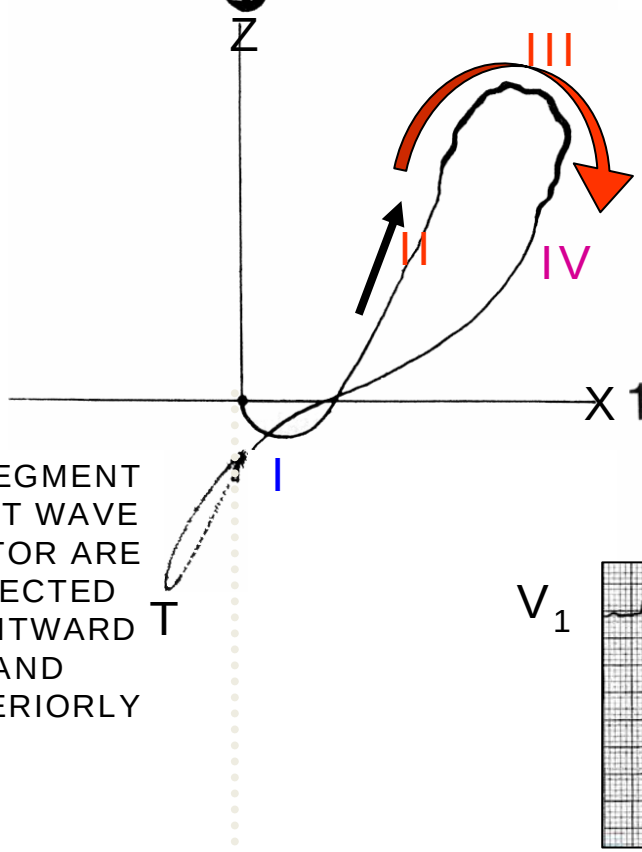
THE FIRST COMPONENT: Right inferior septal activation on the subendocardial region of the anterior papillary tricuspid muscle: **Vector I**: Directed anteriorly, inferiorly and to the left.

THE SECOND COMPONENT: Delayed and anomalous left septal activation: **Vectors II and III**: Directed posteriorly, superiorly and to the left.

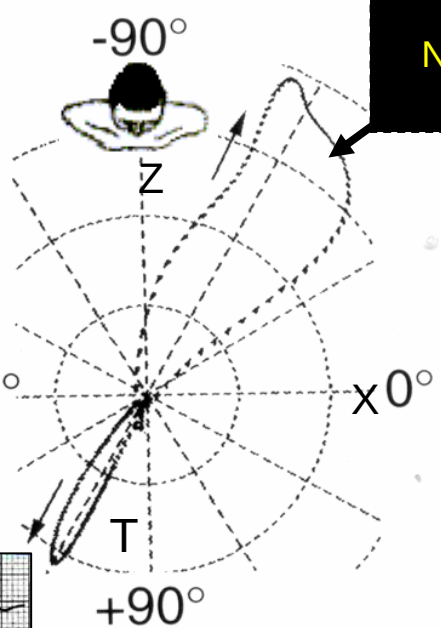
THE THIRD COMPONENT: Delayed and anomalous activation of the free left ventricular wall: **Vector IV**: Directed posteriorly, superiorly and to the left. The last 50ms is made up of activation fronts in the posterolateral wall of the left ventricle.

The QRS loop duration is ≥ 120ms
The QRS loop shape is elongated and narrow
The small short–duration initial 10ms vector directed anteriorly and leftward: **Vector I**
The main body of the QRS loop is inscribed posteriorly and to the left within the range - 90º to – 40 º.
The main body of QRS loop is inscribed clockwise (CW)
The magnitude of the max QRS vector is increased above normal exceeding 2mV.

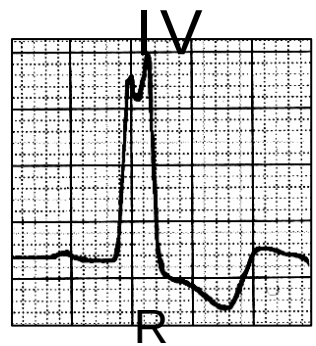


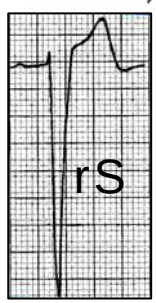


ST SEGMENT AND T WAVE VECTOR ARE DIRECTED RIGHTWARD AND ANTERIORLY



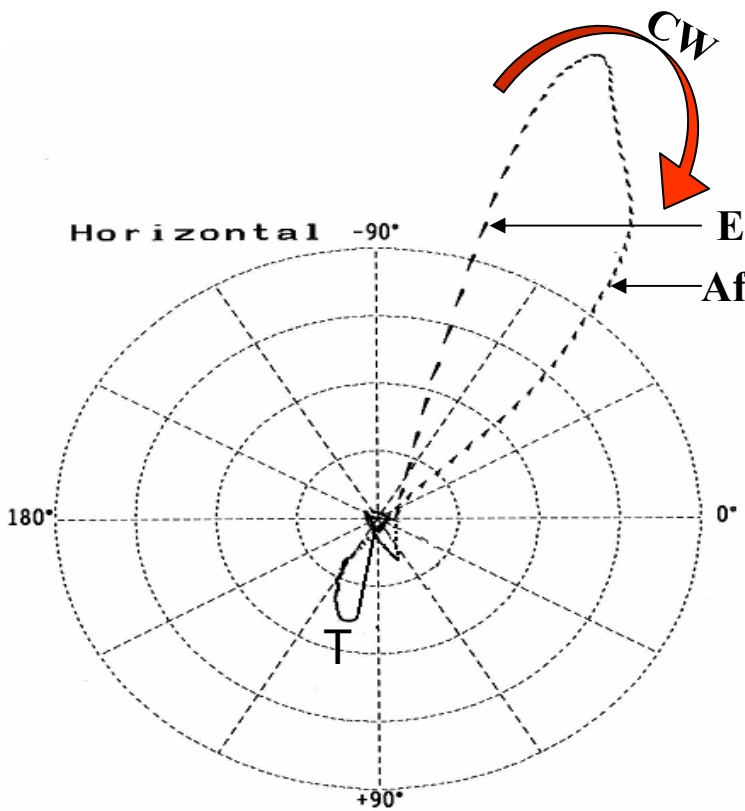
CONDUCTION DELAY NOTED IN THE MID AND TERMINAL PORTION



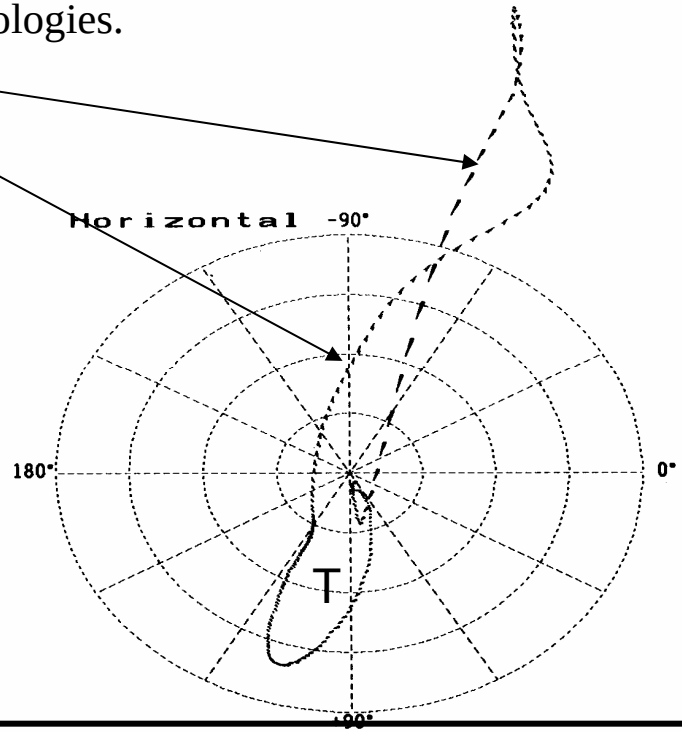


Outline that shows the four activation vectors in LBBB in the horizontal plane. There is an ECG/VCG correlation of the QRS loop and the leads V_1 and V_6. Take notice that the middle final delay of the QRS loop in the HP rotates in a clockwise direction.

Truly LBBB QRS and T loop in HP

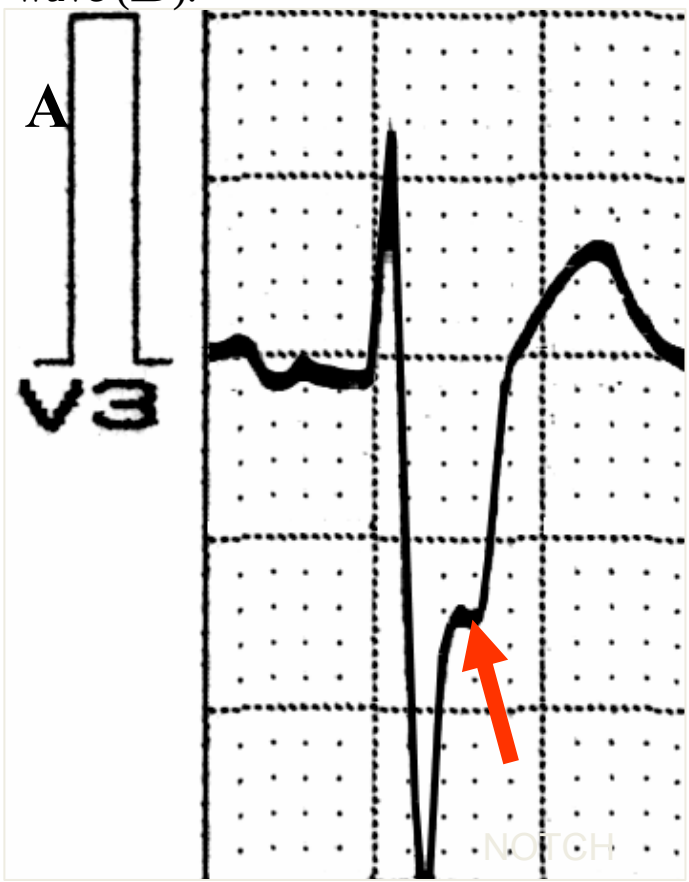


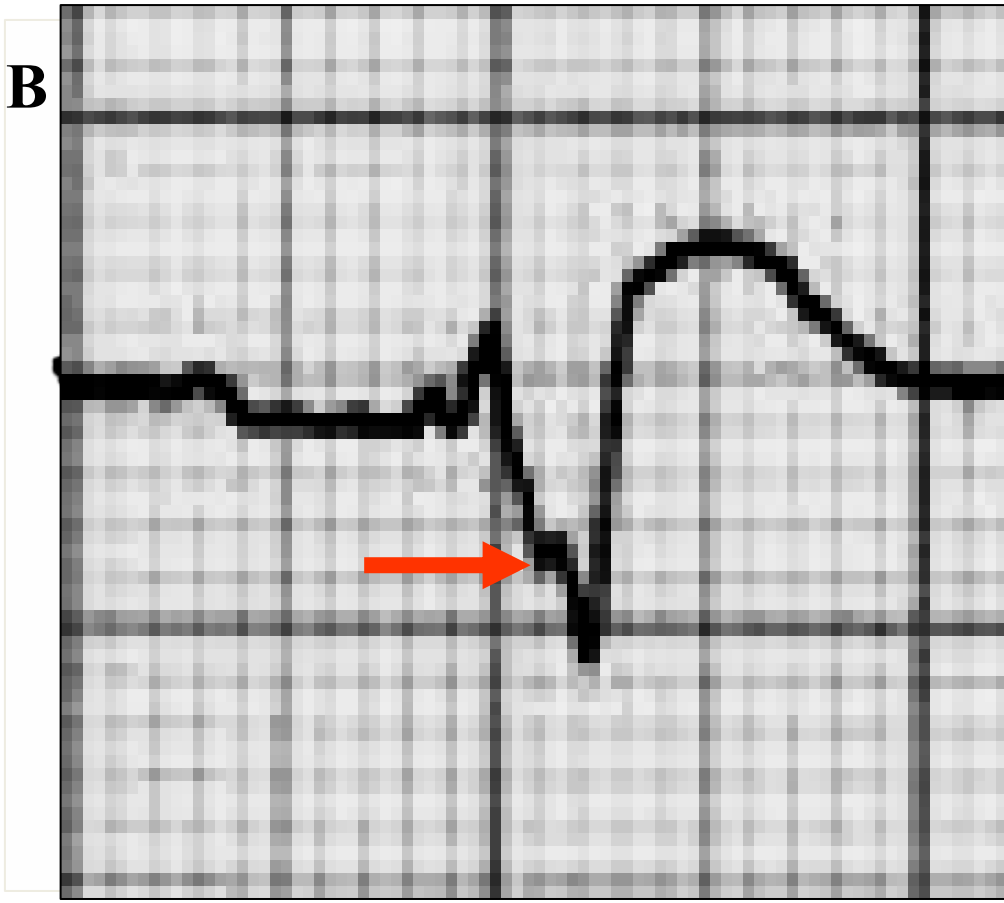
Parietal, intramural, Purkinje or intramyocardial block It is considered a Nonspecific intraventricular conduction disturbance or Nonspecific intraventricular conduction delay non-typical LBBB morphologies.



Main body of QRS loop rotation	**It is inscribed clockwise (CW)**	**It is inscribed counter clock wise rotation or figure in eight**
Afferent limb	**To left related efferent limb**	**To right related efferent limb**
T-Loop shape	**Elongated, asymmetrical and directed rightward and anteriorly**	**Broad and symmetrical**
Cardiac resynchronization therapy (CRT) respond	**Responders**	**Nonresponders.**

The present case is not equal to Cabrera's sign (**A**) (LBBB complicated with MI) because the notch is not in the ascending ram of S wave of V_3 and /or V_4. In this case (**B**) notch is located in descending ramp of S wave (**B**).

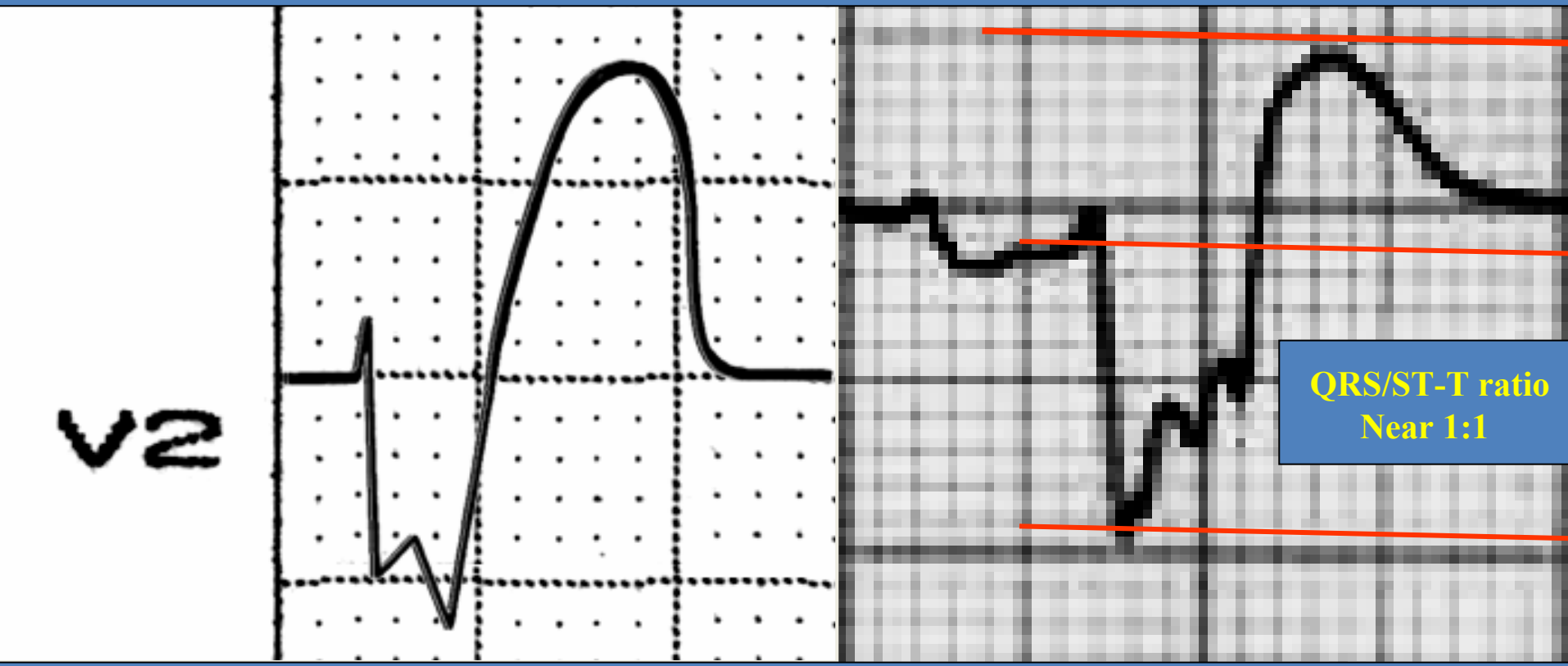




Notch of 50 ms in the ascending ramp of S wave of V_3 and V_4. It is seen more often with MI than without (anterior more often than inferior), and the left axis increased its sensitivity[1;2]

1) **Kindwall KE, Brown JP, Josephson ME.Predictive accuracy of criteria for chronic myocardial infarction in pacing-induced left bundle branch block. Am J Cardiol. 1986; 57:1255-1260.**
2) **Cabrera E, Friedland C. Wave of ventricular activation in left bundle branch block with infarct: a new electrocardiographic sign. Gac Med Mex. 1953; 83:273-280.**

QRS/ST-T ratio in lead V_2 or V_3

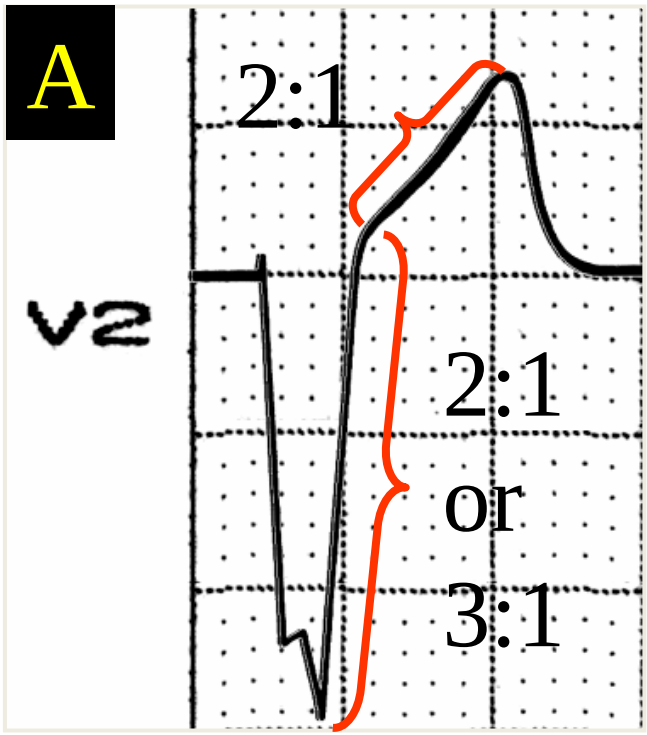


Diminution of QRS/ST-T ratio in lead V_2 or V_3: In uncomplicated LBBB, the ratio of QRS voltage to the ST segment voltage is always greater than 1. Usually 2:1 or 3:1 in V_2 or V_3 lead(1). During AMI the elevation of ST segment with concomitant eventual reduction in the QRS voltage results in a QRS/ST-T ratio near to 1:1 such as the present case without acute MI.

1. **Schamroth L. The Electrocardiology of Coronary Artery Disease Myocardial Infarction Associated with Left Bundle Branch Block 1975; Chapter 10 pp: 93 Blackwell Scientific Publications.**

ECG CRITERIA IN LBBB COMPLICATED WITH AMI

UNCOMPLICATED LBBB



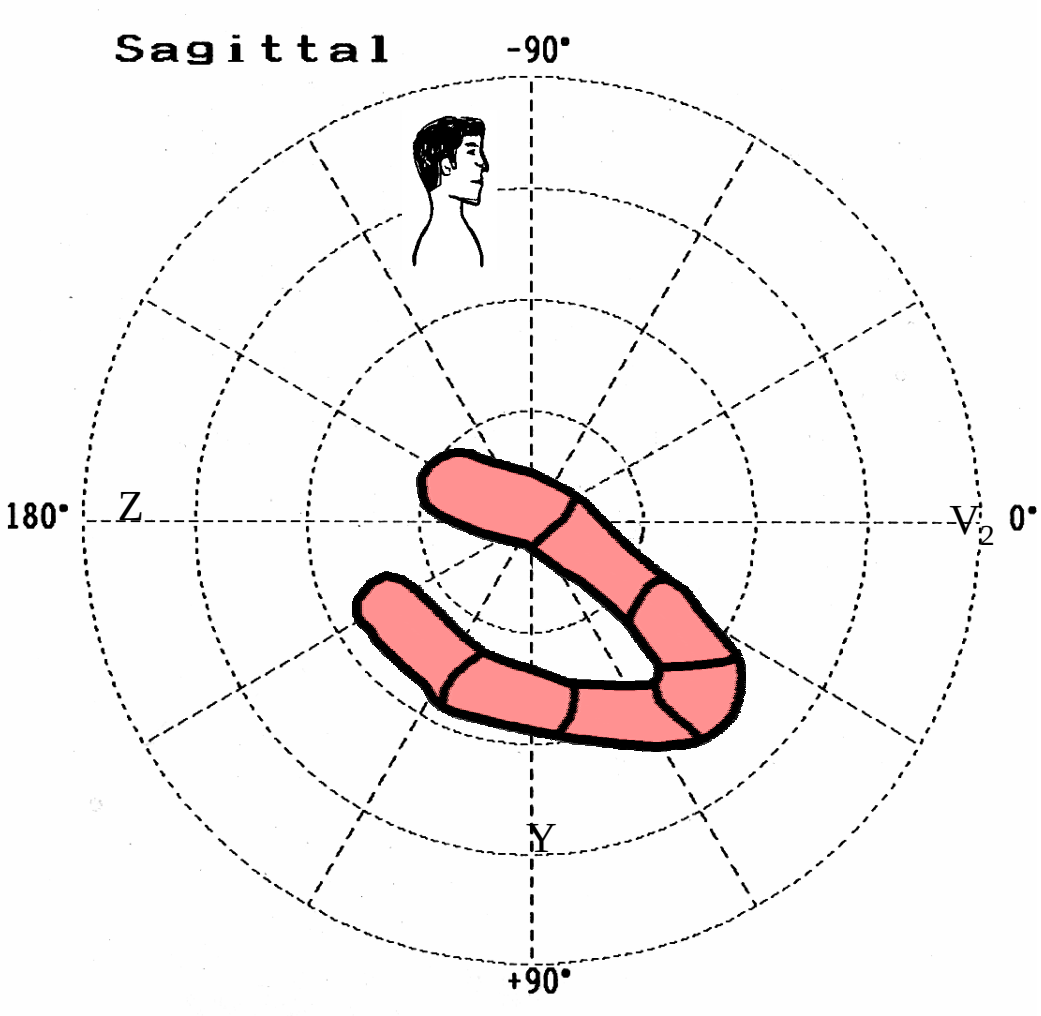
A: Ratio of $^{QRS}/_{ST-T}$ amplitude, 2:1. ST upwardly concave.

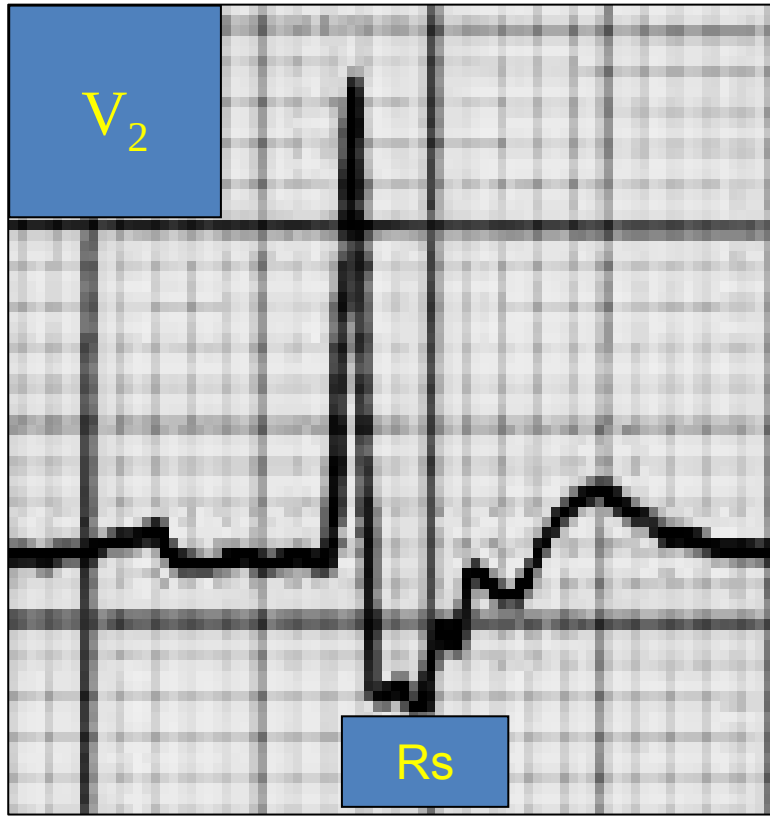
LBBB COMPLICATED
WITH ANTERIOR INACTIVE AREA

B: Ratio of $^{QRS}/_{ST-T}$ amplitude 1:1. ST upwardly convex.

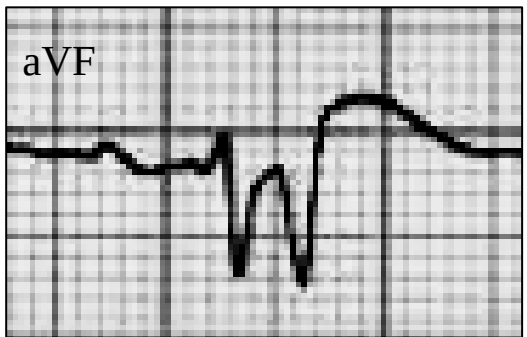
Diminution of $^{QRS}/_{ST-T}$ ratio in lead V2or V3: In uncomplicated LBBB, the ratio of QRS voltage to the ST segment voltage is always greater than 1. Usually 2:1 or 3:1 in V2 lead[1]. During AMI the elevation of ST segment with concomitant eventual reduction in the QRS voltage results in a $^{QRS}/_{ST-T}$ ratio near to 1:1.

1) **Schamroth L. The Electrocardiology of Coronary Artery Disease Myocardial Infarction Associated with Left Bundle Branch Block 1975; Chapter 10 pp: 93 Blackwell Scientific Publications**





This QRS pattern (Rs) in V_2 is not compatible with LBBB



According to my understanding this isolated prominent R wave observed in V_2 could just explained by three mechanisms that we will proceed explaining

1) ***The "Standard Type" or Standard Masquerading Bundle-Branch Block associate to the severe anterolateral fibrosis (it hides the RBBB in precordial leads) and a high degree left anterior fascicular block (extreme left axis deviation on frontal plane)***
2) ***Sequel of electrically inactive area in the lateral wall(1) (dorsal inactive area in ancient nomenclature). This hypothesis has support in Ecocardiograma's findings which described a significant trabeculation precisely in the lateral wall of the left ventricle***
3) ***Some degree of left septal fascicular block (LSFB) consequence of fibrotic involvement of the third fascicle in the anteroapical endocardium of the left ventricle where the fascicle run. Myocardial fibrosis is related to clinical disease severity and LV systolic dysfunction in isolated LVNC. (2)***

1) Masquerading Bundle-Branch Block

The ECG complex coined since Richman as "masquerading bundle-branch block"(3) to day we know that is essentially a complete RBBB and high degree LAFB, with further modifications of the initial and final QRS vectors, so that standard leads, and at times the left precordial leads, resemble left bundle –branch block(4). Masquerading BBB is not a specific entity but is the result of RBBB with varying combinations of LAFB, intramural left ventricular block, left ventricular enlargement and anterior myocardial infarction/fibrosis.

1. ***Bayés de Luna A, Wagner G, Birnbaum Y, et al. International Society for Holter and Noninvasive Electrocardiography.Circulation. 2006 Oct 17;114:1755-1760.***
2. ***Nucifora G, Aquaro GD, Pingitore A,et al. Myocardial fibrosis in isolated left ventricular non-compaction and its relation to disease severity. Eur J Heart Fail. 2011 Feb;13:170-176.***
3. **Richman JL, *Wolff L. Left bundle branch block masquerading as right bundle branch block. Am Heart J. 1954 Mar; 47: 383-393.***
4. ***Schamroth L, Dekock J. The concept of 'masquerading' bundle-branch block. S Afr Med J. 1975 Mar 15; 49: 399-400.***

Since the pioneer Rosembaum´s et al studies (1;2) we know two ECG types of Masquerading Bundle-branch Block. There are a third type that is the association of both:

I. The "Standard Type" or Standard Masquerading Bundle-Branch Block
II. The "Precordial Type" or Precordial Masquerading Bundle-Branch Block
III. The Standard and Precordial Masquerading Bundle-Branch Block in Association.

I. The "standard type" (*"standard masquerading right bundle-branch block")*

In "*standard masquerading right bundle-branch block* the presence of a high degree left anterior fascicular block (LAFB) obscured totally or partially the diagnosis of right bundle branch block (RBBB) only on frontal plane by abolishing (or becomes very small) the final broad S wave in the leads I and aVL (1). Consequently, the limb leads may resemble left bundle branch-block (LBBB) although the precordial ECG remain typical for CRBBB. The precordial leads reflect the feature of RBBB. (see the Figure of next slide)

Conditions necessary for the presence of standard masquerading right bundle –branch block

1. High degree of left anterior fascicular block
2. Right Bundle-Branch Block(3)
3. Frequent Left Ventricular Enlargement or Hypertrophy (LVE/LVH)
4. Localized block in the left ventricle.
5. Frequent severe fibrosis, or truly massive myocardial infarction mainly in anterior wall.

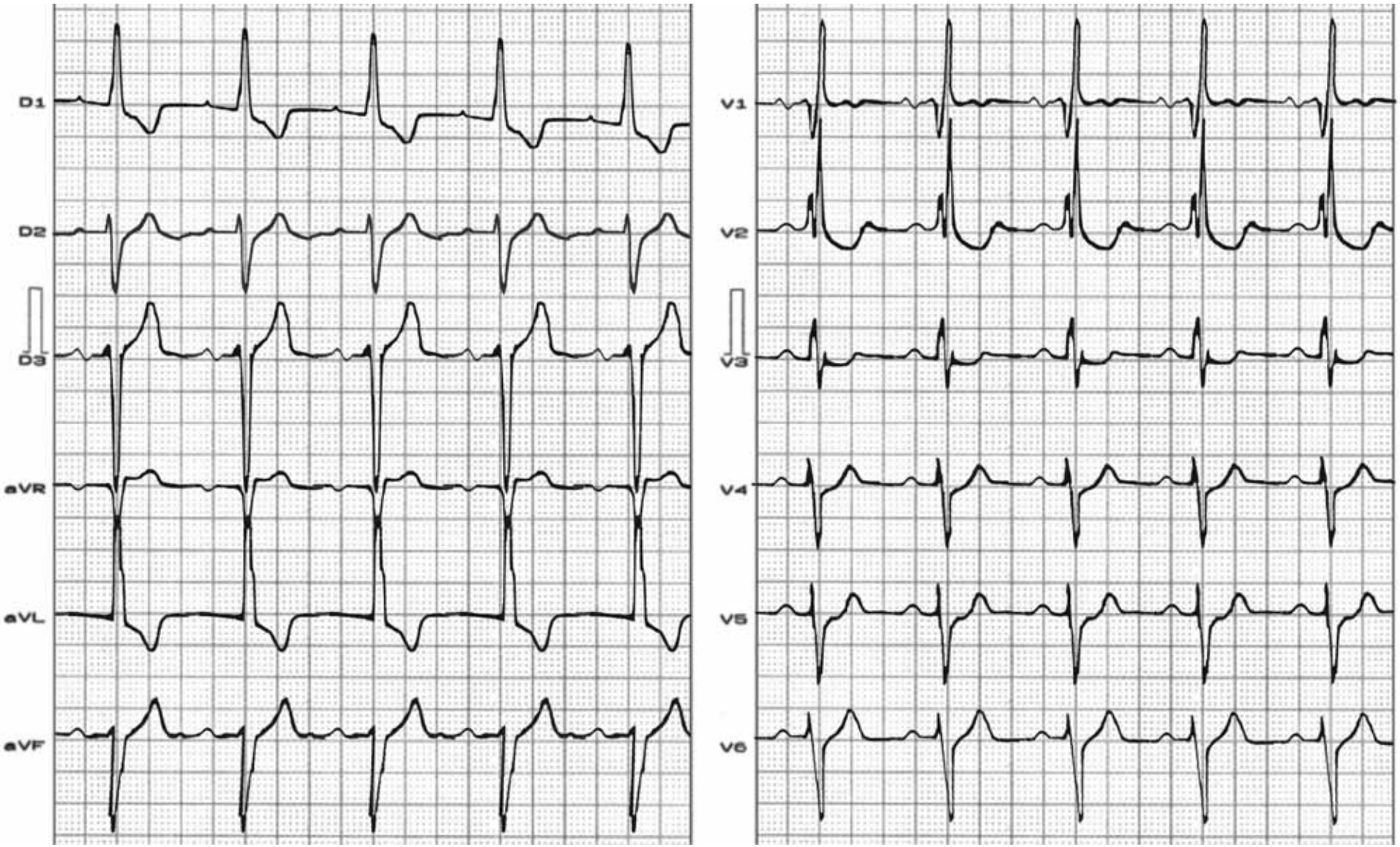
Possible Etiologies

1) Coronary heart disease
2) Long standing systemic hypertension
3) Cardiomyopathies Ex. Chagas myocarditis, Left Ventricular Non-Compaction (LVNC) cardiomyopathy,
4) Lev´s disease.

Prognosis: always poor.

1. **Rosembaum MB,, Elizari MV, Lazzari JO. Los hemibloqueos. Buenos Aires; Paidos 1968.**
2. **Rosenbaum MB, Yesuron J, Lazzari JO, Elizari MV. Left anterior hemiblock obscuring the diagnosis of right bundle branch block. Circulation. 1973 Aug; 48: 298-303.**
3. **Ortega-Carnicer J, Malillos M, Muñoz L, et al. Left anterior hemiblock masking the diagnosis of right bundle branch block. J Electrocardiol. 1986 Jan; 19: 97-98.**

Figure: Typical example of *"Standard Masquerading Right Bundle-branch Block"*



Extreme QRS left axis deviation (SÂQRS -50º), SIII>SII: LAFB. The limb leads show a LBBB-like pattern, but the precordial leads show a RBBB. SIII > 15mm: Type IV Rosembaum LAFB: association of LAFB + LVE or LVH.

II. The precordial type (*"precordial masquerading right bundle-branch block"*

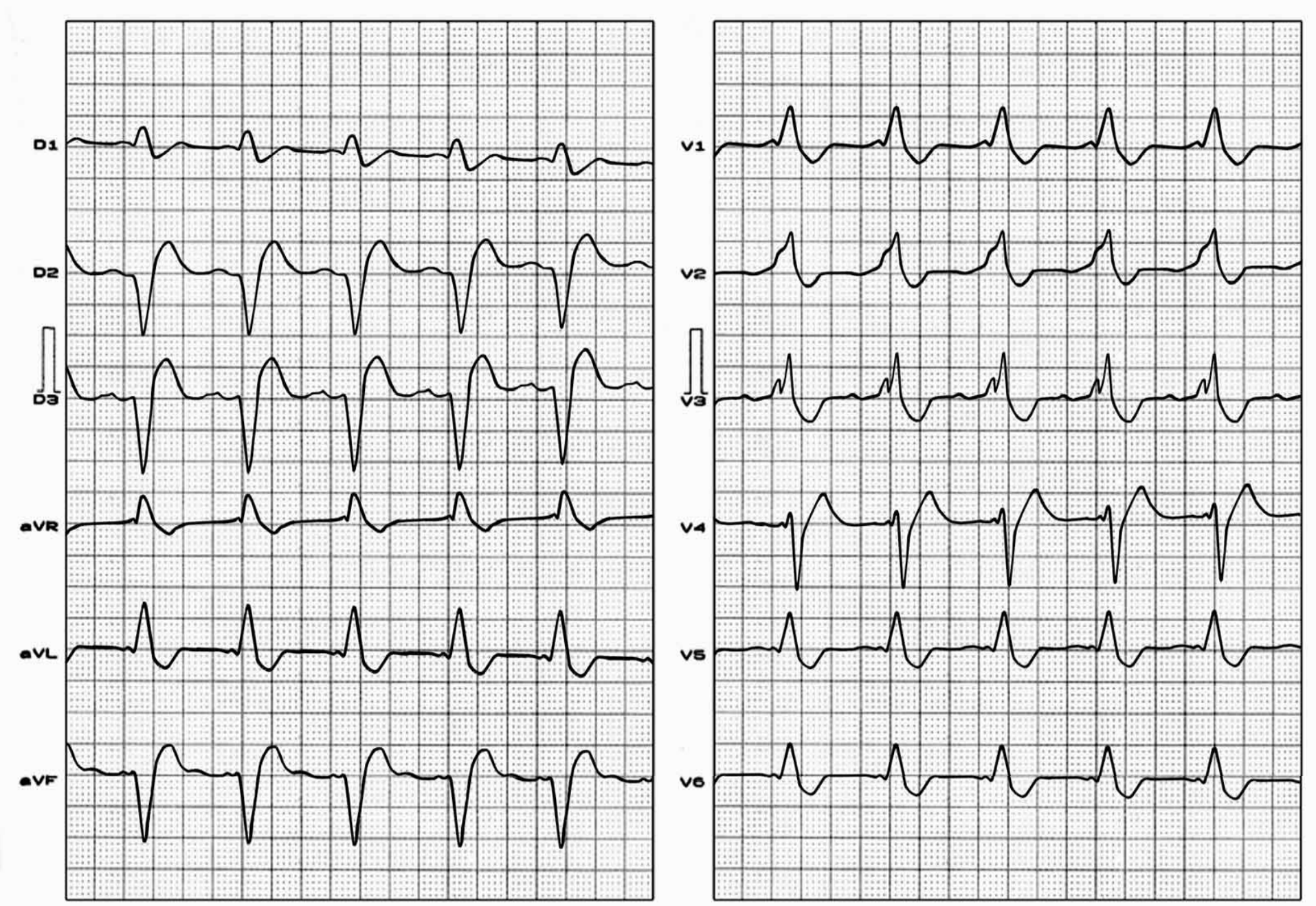
This type shows the pattern of CRBBB in the right precordial leads and complete left branch block pattern (CLBBB) in the left-side precordial leads. This result from CRBBB associated with severe left ventricular hypertrophy (LVH), a localized block in the anterolateral wall of the left ventricle often due to myocardial infarction, and usually LAFB. Presumably, the intramural left ventricular block, together with the LVH or the LAFB, or both, produce predominant leftward forces which tend to cancel out the late rightward forces of the RBBB in the left precordial leads. Finally, masquerading bundle-branch block can be associated with severe and diffuse conduction system disease, and that patients with this finding may require permanent pacemaker implantation, especially if they are symptomatic. (1)

III. The Standard And Precordial Masquerading Bundle-Branch Block In Association

In these cases the limb leads show an apparent Left bundle-branch block pattern with extreme left axis deviation (LAFB) and the precordial leads exhibit the pattern of CRBBB in the right precordial leads and LBBB pattern in left precordial leads V_5-V_6. Additionally, an abnormal Q waves are frequently present on right precordial leads. (see ECG of next slide)

1. **Kowey PR, Koslow M, Marinchak RA Masquerading Bundel-branch block – Electrophysiological correlation J Electrophysiol. 1989; 3:156-159**.

Typical example of Standard And Precordial Masquerading Bundle-Branch Block

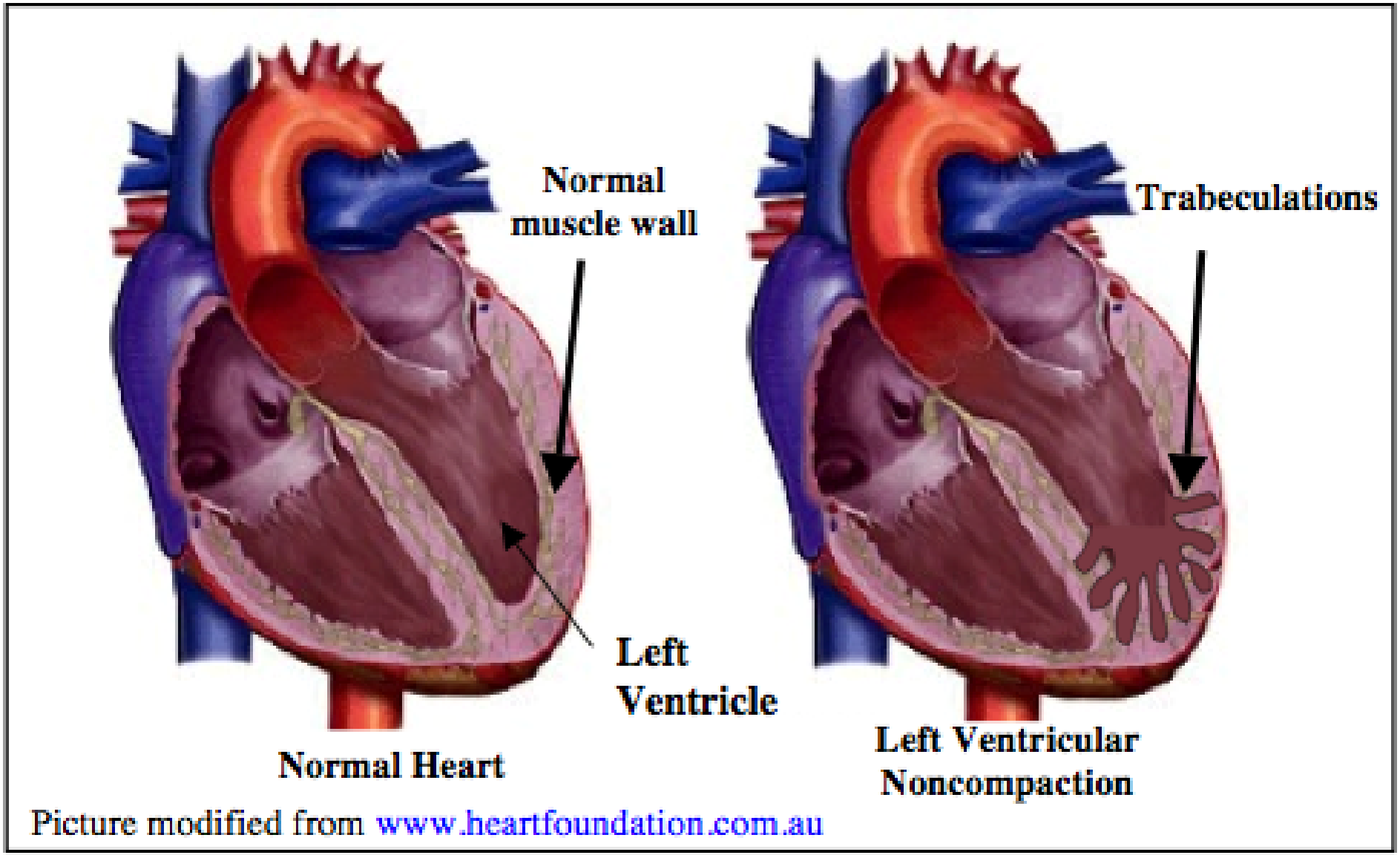


Left Ventricular Non-Compaction (LVNC) cardiomyopathy. **Non-compaction cardiomyopathy** (**NCC**),

Others denominations: Non-compaction of the ventricular myocardium (NVM), spongiform cardiomyopathy
Left Ventricular Non-Compaction (LVNC) cardiomyopathy is a rare congenital cardiomyopathy thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium.

This abnormality is often associated with other congenital cardiac defects, but it is also seen in the absence of other cardiac anomalies.
LVNC, is a rare congenital cardiomyopathy that affects both children and adults.(1) It results from the failure of myocardial development during embryogenesis.(2;3) During development, the majority of the heart muscle is a sponge-like meshwork of interwoven myocardial fibers. As normal development progresses, these trabeculated structures undergo significant compaction that transforms them from spongy to solid. This process is particularly apparent in the ventricles, and particularly so in the LV(LVNC). NCC results when there is failure of this process of compaction. Because the consequence of LVNC is particularly evident in the LV, the condition is also called LVNC..

1. Pignatelli RH, McMahon CJ, Dreyer WJ, *et al* (November 2003). "Clinical characterization of left ventricular noncompaction in children: a relatively common form of cardiomyopathy". *Circulation; 2003;* 108 : 2672–2678.
2. Espinola-Zavaleta, Nilda.; Soto, Elena.; Castellanos, Luis Munoz; Játiva-Chávez, Silvio; Keirns, Candace."Non-compacted Cardiomyopathy: Clinical-Echocardiographic Study".2006; *Cardiovasc Ultrasound* 4 (1): 35.
3. Oechslin, Erwin; Jenni, Rolf (2005) (webpage). *Non-compaction of the Left Ventricular Myocardium - From Clinical Observation to the Discovery of a New Disease*. Archived from the original on 2007-09-29.



Left Ventricular Non-Compaction (LVNC) is characterized by deep trabeculations (finger-like projections) in the muscle wall of the left ventricle. These trabeculations can also occur in the right ventricle. The heart muscle abnormalities occur during the development of the heart in the embryo.

History

NCC was first identified as an isolated condition in 1984 by Engberding and Benber.(1)

These authors reported on a 33 year old female presenting with exertional dyspnea and palpitations. Investigations concluded persistence of myocardial sinusoids (now termed non compaction). Prior to this report, the condition was only reported in association with other cardiac anomalies, namely pulmonary or aortic atresia. Myocardial sinusoids is considered not an accurate term as endothelium lines the intertrabecular recesses.

1. Engberding R, Bender F: Identification of a rare congenital anomaly of the myocardium by two-dimensional echocardiography: Persistence of isolated myocardial sinusoids. Am J Cardiol 1984 Jun 1;53:1733-4

Genetics

Families with LVNC have been shown to pass the disease on in two different ways, via autosomal dominant or x-linked inheritance.

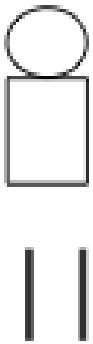
Autosomal Dominant Inheritance.
This means that an affected person has a 1 in 2 (50%) chance of passing the gene alteration on to children and males and females are affected equally. Most inherited heart diseases are passed on in this fashion.

X-Linked Inheritance.
This type of inheritance has so far only been observed in a small group of patients who develop LVNC during childhood. In this case the gene alteration exists in a gene located on the X chromosome (sex chromosomes determine gender, i.e. females XX, males XY). Therefore, a father with LVNC has a 100% chance of passing the gene alteration on to his daughters but no chance of passing it on to his sons. Alternatively, an affected mother has a 50% chance of passing the gene alteration on to both daughters and sons. A female who inherits the gene alteration may have less chance of actually developing the disease (i.e. may never develop the trabeculations) however can still pass it on to her children.
Rarely, a person may be the first in the family to have the mutation. In this case, brothers and sisters of that person are not likely to develop the disease, however children of the affected person are at risk of inheriting the gene mutation.
Genetic testing involves looking for a mistake in the genes known to cause LVNC. At present, less than five genes have been identified to cause this disease and these genes are poorly understood.
LVNC and to understand how these gene abnormalities cause heart disease. In addition to the genetic studies, clinical and family information is also being collected to help understand more about this disease.

AUTOSOMAL DOMINANT

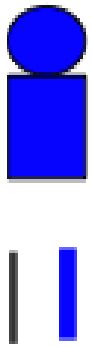
For each pregnancy there is a **1 in 2** chance the child will inherit the gene alteration

Unaffected Parent



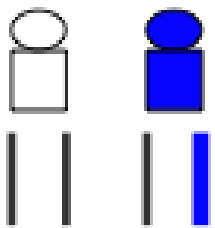
The child is guaranteed to get an unaffected gene from the parent who does not have LVNC

Parent with LVNC



However, they will either inherit the unaffected copy or the altered copy from the affected parent.

50% chance of having a child without LVNC



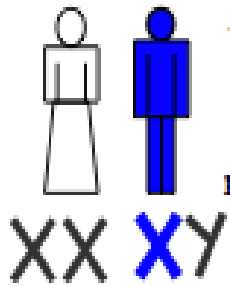
50% chance of having a child with LVNC

| - Unaffected copy **▮ - Altered/affected copy causing LVNC**

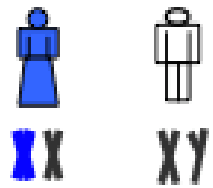
X-LINKED

The chance that a child will be affected depends on which parent has the disease. Males more often are affected

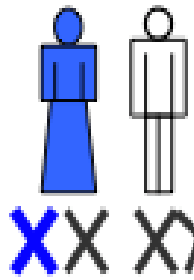
Father with LVNC



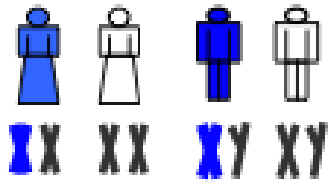
All daughters will inherit the affected X, however may not develop the disease. The father is only able to pass on his Y to his sons



Mother with LVNC



An affected mother has a 50% chance of passing on her affected X to both her sons and daughters. Again, her daughters may not develop the disease



The gene mutation is carried on the X chromosome. Females have two X's while males have an X and a Y

Epidemiology

Due to its recent establishment as a diagnosis, and it being unclassified as a cardiomyopathy according to the WHO, it is not fully understood how common the condition is. Some reports suggest that it is in the order of 0.12 cases per 100,000. The low number of reported cases though is due to the lack of any large population studies into the disease and have been based primarily upon patients suffering from advanced heart failure. A similar situation occurred with Hypertrophic cardiomyopathy which was initially considered very rare; however is now thought to occur in one in every 500 people in the population.
Again due to this condition being established as a diagnosis recently, there are ongoing discussions as to its nature, and to various points such as the ratio of compacted to non-compacted at different age stages. However it is universally understood that LVNC will be characterized anatomically by *deep trabeculations in the ventricular wall, which define recesses communicating with the main ventricular chamber. Major clinical correlates include systolic and diastolic dysfunction, associated at times with systemic embolic events.*(1)

1. The Cardiomyopathy Association (2007-07-23). "LV Non-compaction" (website). http://www.cardiomyopathy.org/index.php?id=274. Retrieved 2007-07-23.

Symptoms

Symptoms range greatly in severity. Most are a result of a poor pumping performance by the heart (congestive heart failure), arrhythmias, and systemic thromboemboli. The disease can be associated with other problems with the heart and the body. Subjects' symptoms from LVNC range widely. It is possible to be diagnosed with the condition, yet not to suffer from any of the symptoms associated with heart disease. Likewise it possible to suffer from severe HF which even though the condition is present from birth, may only manifest itself later in life.

Differences in symptoms between adults and children are also prevalent with adults more likely to suffer from HF and children from depression of systolic function.

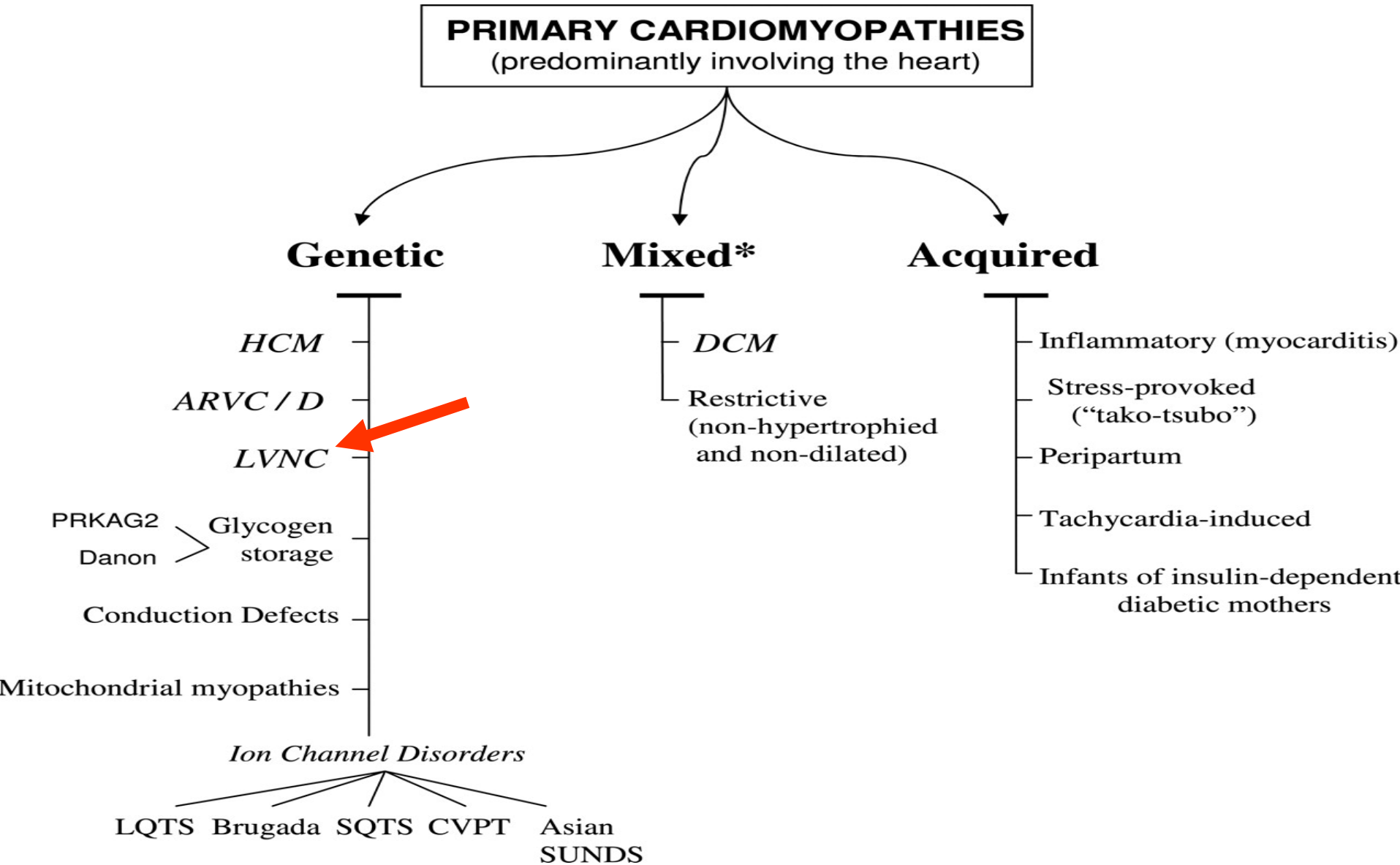
The clinical manifestations include HF signs, ventricular arrhythmias and cardioembolic events.

Main manifestation include(1)

1. Breathlessness
2. Fatigue
3. Swelling of the ankles
4. Limited physical capacity and exercise intolerance
5. Two conditions though that are more prevalent in LVNC are: tachyarrhythmia which can lead to Sudden Cardiac Death and clotting of the blood in the heart

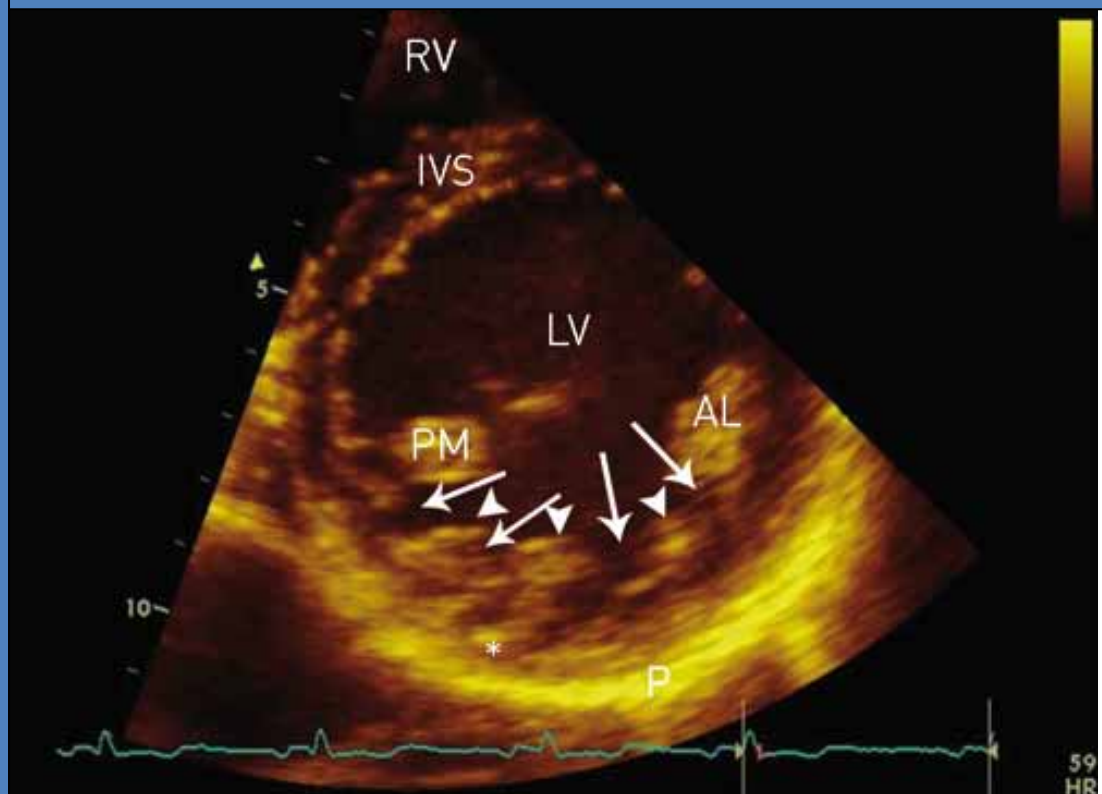
1. Bottio T, Farina D, Piccoli P, et al. Massive mitral and pulmonary valve incompetence in a patient with left ventricular, non-compacted myocardium. J Heart Valve Dis. 2007 Jan;16:93-95.

Primary cardiomyopathies in which the clinically relevant disease processes solely or predominantly involve the myocardium. The conditions have been segregated according to their genetic or nongenetic etiologies. *Predominantly nongenetic; familial disease with a genetic origin has been reported in a minority of cases.



Diagnosis

Trabeculation of the ventricles is normal, as are prominent, discrete muscular bundles greater than 2mm. In non compaction there is excessively prominent trabeculations. The characteristic echocardiographic findings consist of multiple, prominent myocardial trabeculations and deep intertrabecular recesses communicating with the LV cavity. The disease typically involves the LV myocardium, but right ventricular involvement is not uncommon. Echocardiographic findings are important clues for the diagnosis. Chin, et al. .(1) described echocardiographic method to distinguish non compaction for normal trabeculation. They described a ratio of the distance from the trough and peak, of the trabeculations, to the epicardial surfaceNon compaction is diagnosed when the trabeculations are more than twice the thickness of the underlying ventricular wall.



Transthoracic echocardiography, parasternal short axis view of the LV. The inferolateral wall of the LV with prominent trabeculae (arrowheads) and deep intertrabecular recesses (arrows). The normally developed epicardial myocardial layer (star) in the region is notably thinner than normal, when compared with the thickness of IVS.
AL = anterolateral papillary muscle, IVS = interventricular septum, LV = left ventricle, RV = right ventricle, P = pericardium, PM = posteromedial papillary muscle

1. Chin TK, Perloff JK, Williams RG, et al: Isolated noncompaction of left ventricular myocardium. A study of eight cases. Circulation 1990 Aug;82:507-513

LVNC was diagnosed when 3 criteria were established:

1) **The presence of multiple echocardiographic trabeculations**

2) **Multiple deep intertrabecular recesses communicating with the ventricular cavity, as demonstrated by color Doppler imaging and the recesses demonstrated in the apical or middle portion of the ventricle, and**
3) **Layered structure of the endocardium with a noncompacted to compacted ratio >1.4(1)**

LVNC is underdiagnosed as a result of lack of knowledge about the disease and screening of relatives of those presenting with LVNC (2). Though echocardiography has historically been the diagnostic test of choice, MR and more recently CT have been shown to be complimentary or superior in detecting the characteristic, 2-layered myocardium (3).

MR offers high quality images, preventing misdiagnosis of the characteristic trabeculations as apical hypertrophic cardiomyopathy or other cardiac pathology. It allows for acquisition of images in any plane without limitations of acoustic windows and has been utilized to diagnose cases that went undetected by echocardiographic exam

1. Jenni R, Oechslin E, Schneider J, et al. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart.2001 Dec;86:666-671.
2. Goo HW and Park IS. Left Ventricular noncompaction in an infant: use of non-ECG-gated cardiac CT. Pediatric Radiology 2007;37:217-220.
3. Amir, O et al . The Value of Cardiac Magnetic Resonance Imaging in the Diagnosis of Isolated Non-Compaction of the Left Ventricle. Israel Medical Association Journal 2009;11:313-314.

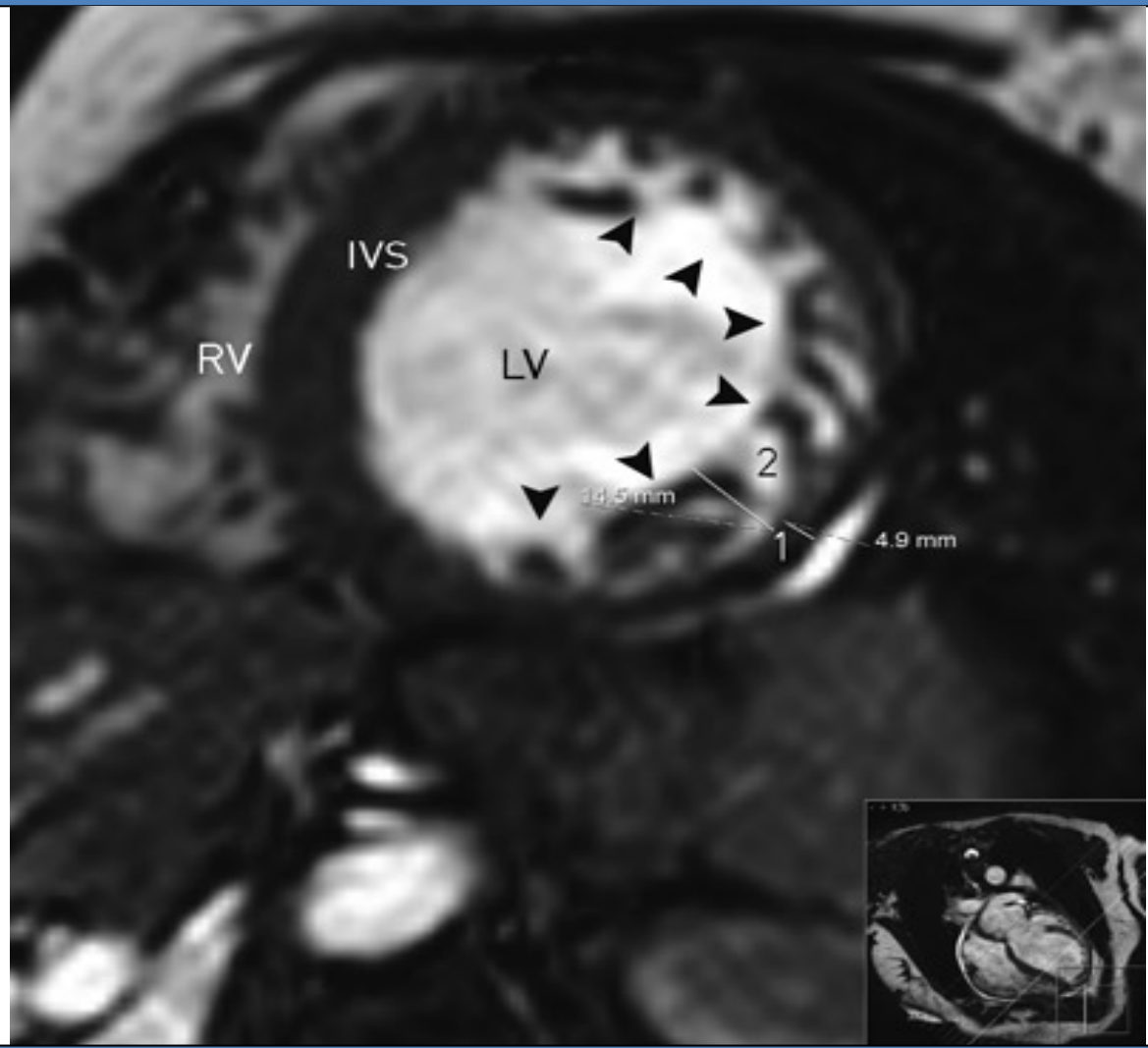
. MR has been found to be diagnostic of LVNC at a ratio of noncompacted to compacted myocardium > 2.3 at the end of diastole, with sensitivities and specificities of 84 and 99% (1). Furthermore, MR delayed hyperenhancement correlates with degree of wall motion abnormality and has been found to be a positive independent predictor of LVEF and disease severity (2).

Others have used the ratio of 2.3 as the basis for CT diagnosis of LVNC (3;4), finding a good correlation to MR results.

CT is advantageous over other modalities in its ability to exclude anomalies of the coronary vasculature which have been described with LVNC (6). This modality further demonstrates a high spatial and temporal resolution with quick acquisition of images as compared to MR (7).

1. **Peterson SE et al. Left Ventricular Noncompaction: Insights from Cardiovascular Magnetic Resonance Imaging. Journal of the American College of Cardiology 2005;46:101-105.**
2. **Dodd, JD et al. Quantification of Left Ventricular Noncompaction and Trabecular Delayed Hyperenhancementwith Cardiac MRI: Correlation with Clinical Severity. American Journal of Roentgenology 2007;189:974-980.**
3. **Conces, Jr., DJ et al. Noncompation of the Ventricular Myocardium: CT Appearance. American Journal of Roentgenology 1991;156:717-718.**
4. **Goo HW, and Park IS. Left Ventricular Noncompaction in an Infant: Use of non-ECG-Gated Cardiac CT. Pediatr Radiol 2007;37:217-220.**
5. **Bladt O et al. Isolated Noncompaction of Ventricular Myocardium. Diagnosis with Multidetector Computed Tomography. Journal Belge de Radiologie - Belgisch Tijdschrift voor Radiologi 2008;91:153-154.**
6. **Hamamichi, Y et al. "Isolated noncompaction of the ventricular myocardium: Ultrafast computed tomography and magntic resonance imaging." The International Journal of Cardiovascular Imaging (2001) 17: 305-314.**
7. **Ito H and Dajani KA. A case with noncompactionof the left ventricular myocardium detected by 64-slice multidetector computed tomography." Journal of Thoracic Imaging (2009) 24:38-40.**

Noncompaction of the left-ventricular myocardium magnetic resonance imaging



Magnetic resonance tomography of the heart. End-diastolic short axis view of the LV and RV. The lateral wall and adjacent segments of the inferior and anterior wall (arrowheads) with noncompaction changes, including a thinner epicardial layer (measurement 1:4.9 mm) and a thicker trabecularized endocardial layer (measurement 2: 14.5 mm). End-diastolic thickness of the inner layer is 3.0 times that of the outer layer. LV = left ventricle, RV = right ventricle

Complications

The presence of LNCC can also lead to other complications around the heart and elsewhere in the body. These are not *necessarily* common complications and no paper has yet commented on how frequently these complicationcs occur with LNCC as well.

Cardiac

1. **Abnormalities of the origin of the left coronary artery**
2. **Pulmonary atresia**
3. **Stenosis**
4. **Right or Left ventricle obstruction**
5. **Hypoplastic left ventricle**
6. **Mitral regurgitation**

Neuromuscular (Pertaining to both nerves and muscles)

1. **Becker's muscular dystrophy**
2. **Mitochondrial myopathy**
3. **Polyneuropathy and metabolic myopathy**

Genetic related

1. **Emery-Dreifuss muscular dystrophy**
2. **Myotubular cardiomyopathy**
3. **Barth syndrome**

Differential diagnosis

1. **Dilated cardiomyopathy**
2. **Congenital heart disease**
3. **Ischemic heart disease**
4. **Disease of the heart valves**
5. **Dilated phase hypertensive cardiomyopathy**
6. **Restrictive cardiomyopathy.**

Advances in medical imaging equipment have made it easier to diagnose the condition, particularly with the wider use of MRIs

Prognosis

Due to LVNC being a relatively new disease, its impact on human life expectancy is not very well understood. In a 2005 study which documented the long term follow up of 34 patients with LVNC, 35% had died at the age of 42 +/- 40 months with a further 12% having to undergo a heart transplant due to HF. However, this study was based upon symptomatic patients referred to a tertiary care center, and so were suffering from more severe forms of LVNC than might be found typically in the population. As LVNC is a genetic disease, immediate family members are being tested as a precaution which is turning up more supposedly healthy people with LVNC who are asymptomatic. The long term prognosis for these people is currently unknown.

Management

Various types of management of care that have been used for various types of LVNC. These are similar to management programs for other types of cardiomyopathies which include the use of ACE inhibitors, beta blockers and aspirin therapy to relieve the pressure on the heart, surgical options such as the installation of pacemaker is also an option for those thought to be at a high risk of arrhythmia problems.

In severe cases, where LVNC has led to heart failure, with resulting surgical treatment including a heart valve operation, or a heart transplant.

Main features

1. ***LVNC may cause heart failure, systemic thromboembolism, serious ventricular arrhythmias and death.***
2. ***LVNC is caused by incomplete endomyocardial development of the LV in early fetal life.***
3. ***LVNC is an Autosomal Dominant Inheritance or X-Linked Inheritance disease.***
4. ***LVNC is actually considered a primary cardiomyopathy***
5. ***LVNC patients, the presence of f-QRS has significant prognostic value and may provide a valid method of risk stratification. The ECG may reveal abnormalities but these are usually non-specific,***
6. ***LVNC is diagnosed by two-dimensional echocardiography and/or cardiac MRI.***
7. ***CT is advantageous over other modalities in its ability to exclude anomalies of the coronary vasculature which have been described with LVNC. This modality further demonstrates a high spatial and temporal resolution with quick acquisition of images as compared to MR***
8. ***There is reason to believe that LVNC is more common than previously assumed.***