CAUCASIAN WOMAN OF AVEARGE AGE WITH PROGRESSIVE MUSCULAR DISEASE FROM CHILD, RECENT CONGESTIVE HEART FAILURE COMPLICATED WITH ACUTE ATRIAL FIBRILLATION AND SYSTEMIC EMBOLISM

MULHER DE MÉDIA IDADE COM DOENÇA PROGRESSIVA MUSCULAR DEBILITANTE DESDE CRIANÇA E RECENTE QUADRO DE INSUFICIÊNCIA CARDIACA CONGESTIVA COMPLICADO COM FIBRILAÇÃO ATRIAL AGUDA E EMBOLIA SISTÊMICA

Case report from Raimundo Barbosa Barros MD
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Final commentaries Andrés Ricardo Pérez-Riera M.D. Ph.D.
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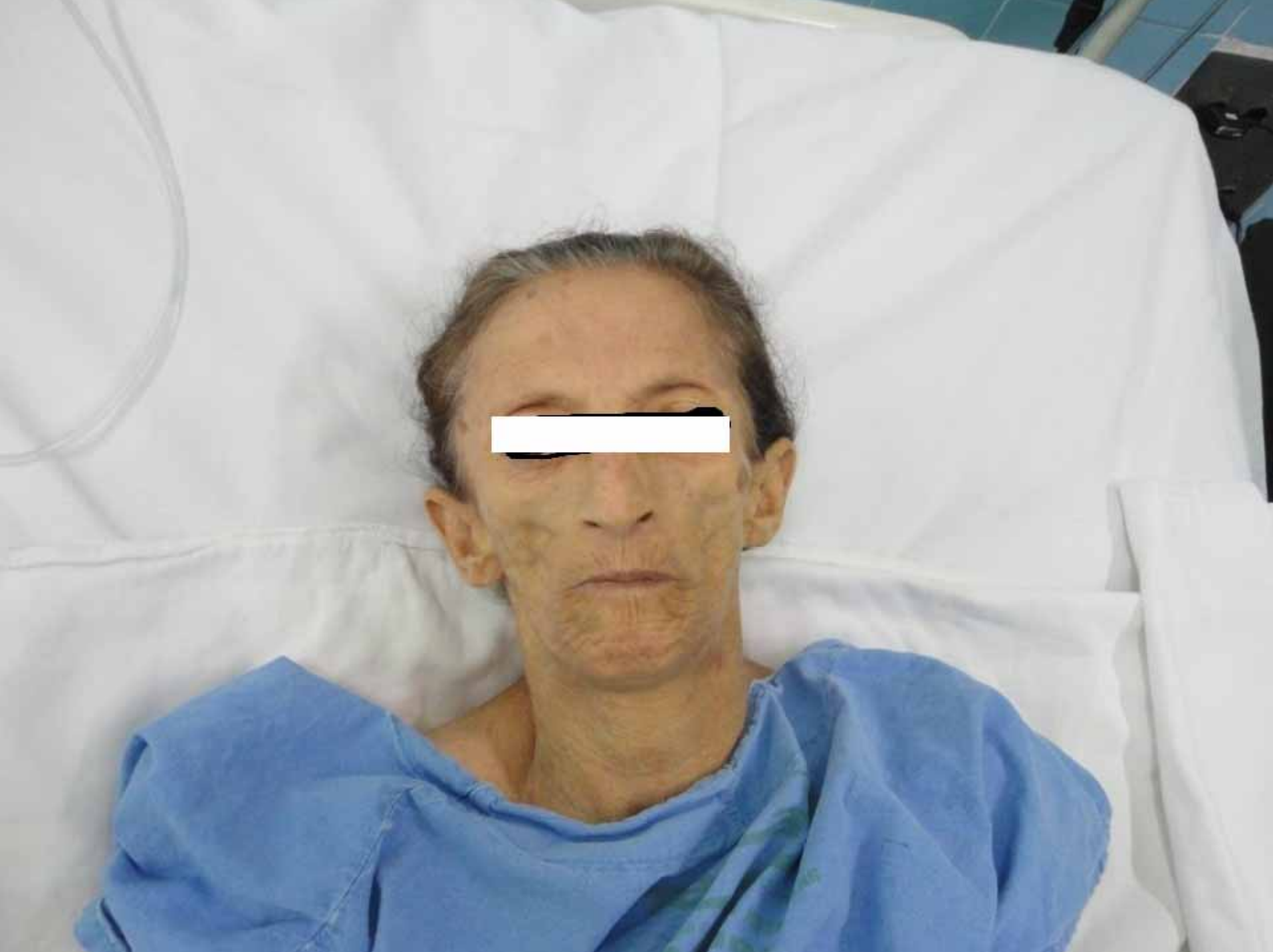
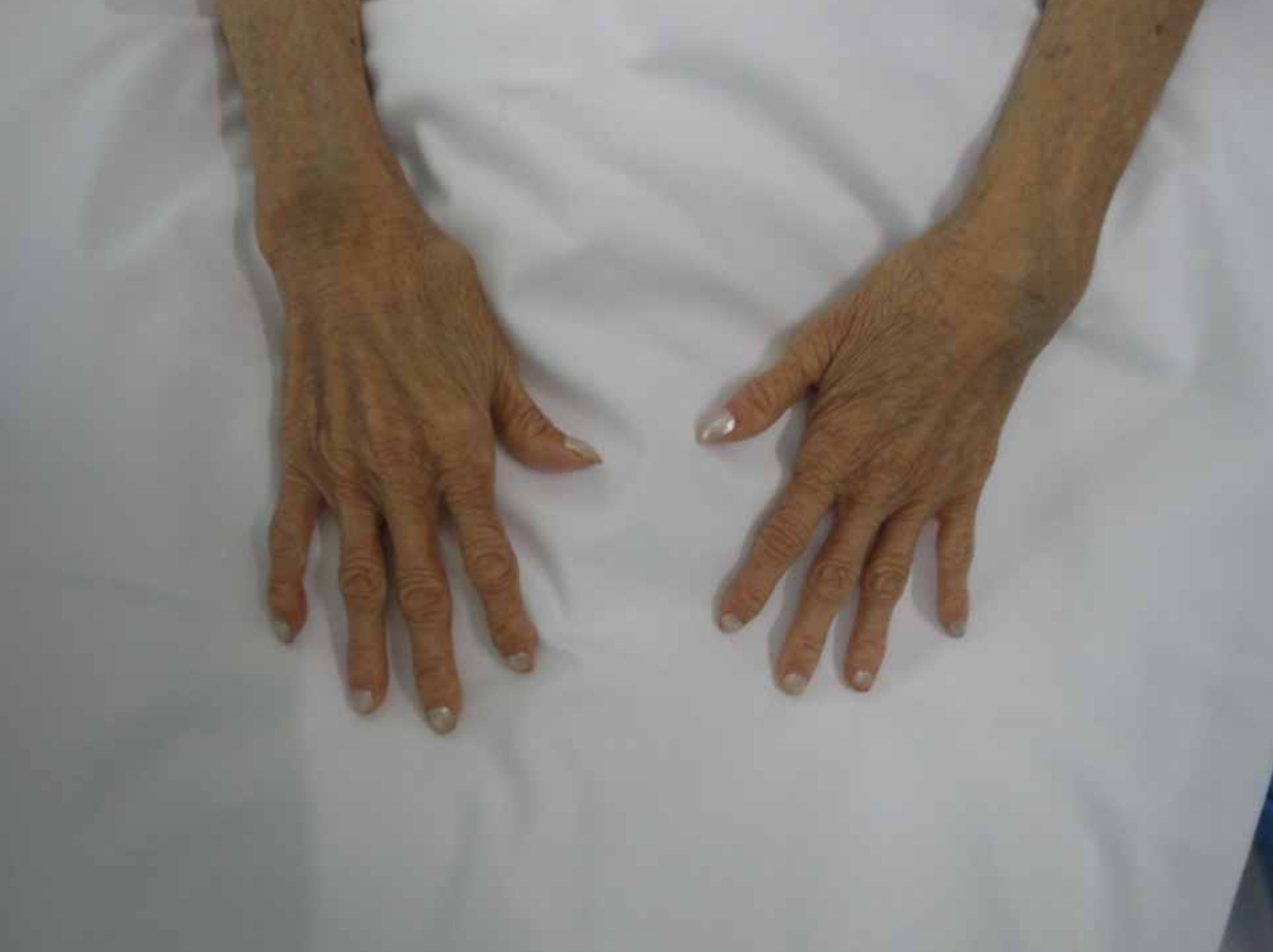
Qual é o diagnóstico clínico-eletrocardiográfico desta paciente?
Infelizmente não tenho muitos detalhes pois a paciente foi internada faz poucas hóras na nossa enfermaria,entretanto penso que o ECG é de grande valor.

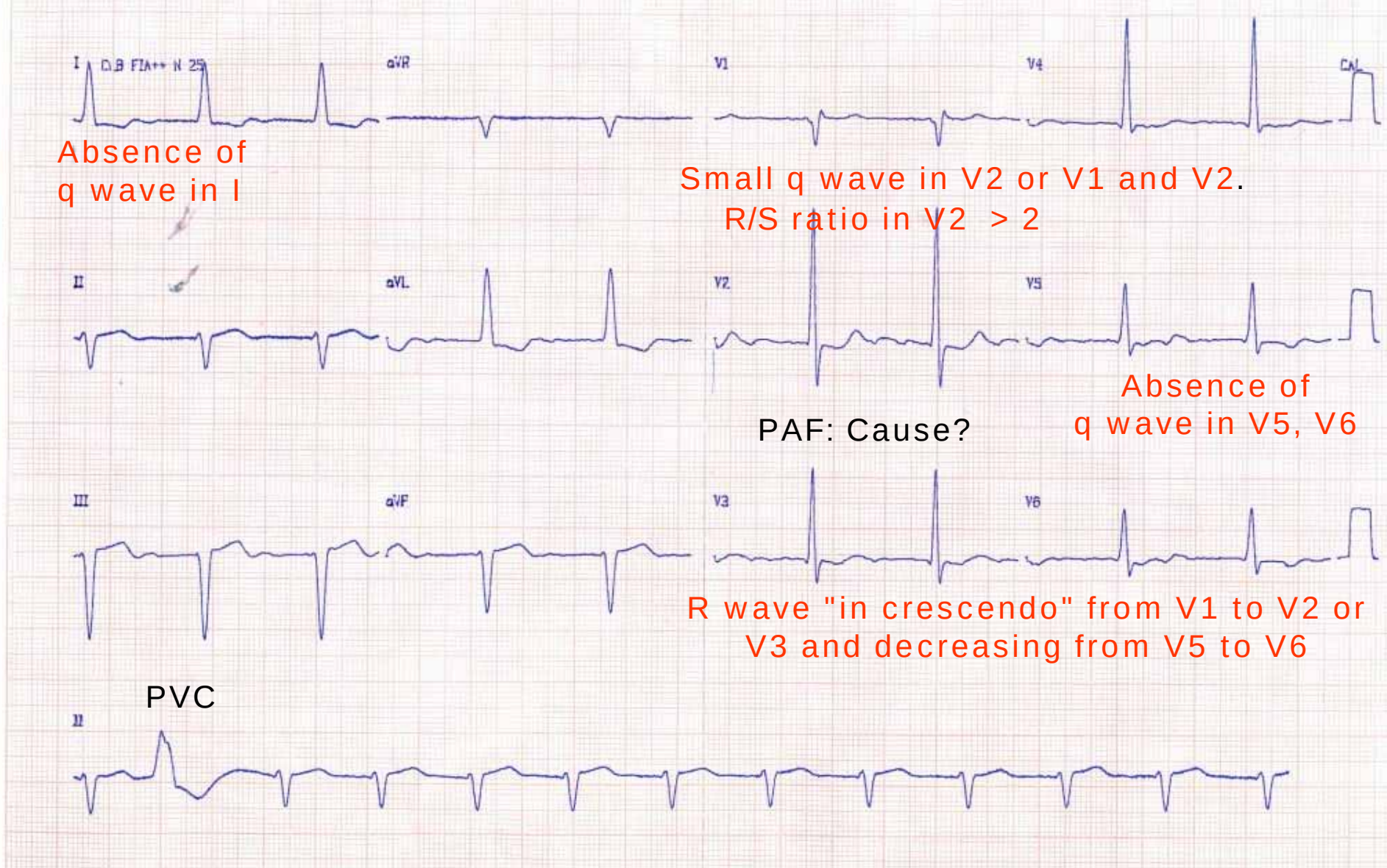
Trata-se de uma mulher branca de 52 anos admitida com quadro de insuficiência cardíaca, complicado com fibrilação atrial aguda e embolia periférica de membors inferiores; Segundo os familiares a sua doença se remonta desde a terna infância (a partir dos 5 anos) e sendo caracterizada por fraqueza muscular progressiva e nos últimos anos dispnéia progressiva.

What the clinical-electrocardiographic diagnosis of this patient?
Unfortunately I do not have much detail because the patient was hospitalized a few hours ago in our ward, but I think the ECG has a great value diagnosis.

It is a Caucasian woman, 52 years admitted with picture of congestive heart failure complicated by acute atrial fibrillation and peripheral systemic embolism in the lower limbs.
According her family the illness start with 5 years old and was characterized by progressive muscle weakness and in the last years progressive dyspnoea and fatigue.







Extreme left axis deviation on frontal plane: -45º, SIII>SII, final s waves in left precordial lead: Left Anterior Fascicular Block (LABF). Normal QRS duration or with a discrete increase (up to 110 ms). Absence of initial q waves in left leads by absence of 1AM QRS vector(first septal 10 ms QRS vector dependent of left septal fascicle

Colleagues commentaries

Estimado Maestro dos consideraciones una clinica y otra electrocardiográfica. Usted a enviado las fotos asi que me permito ese desiz.

1. **Diagnóstico clinico: esclerodermia. Con esa facies sin ver la foto de las manos seria mi primera impresión diagnóstica**
2. **ECG: ritmo regular 75 por minuto, no logro visualizar las ondas P en DI y V5 impresiona inscribirse con PR de 0,60 seg. Eje desviado a la izquierda por bloqueo del fasciculo anterior izquierdo. Ondas R altas en V2 y V3 probablemte por hipertrofia septal. Signos de sobrecarga VI.**

En la esclerodermia puede hallarse afectación musculocutánea con debilidad proximal progresiva y afectación cardiaca, con desarrollo de miocardiopatia hipertrofia.

Saludos
Martin Ibarrola

Bloqueo aurículo-ventricular completo como signo de afección cardíaca en paciente con esclerodermia omero Requena, JM; Bureo Dacal, JC Publicado en Semergen. 2007;33:549-51. - vol.33 núm 10

http://www.elsevier.es/es/revistas/semergen-medicina-familia-40/bloqueo-auriculo-ventricular-completo-como-signo-afeccion-cardiaca-13113218-cartas-al-director-2007

Severe cardiac involvement in children with systemic sclerosis and myositis. Pierre Quartier http://www.jrheum.org/content/29/8/1767.short

Dear Master Andrés,

Two reflections, one clinical and another electrocardiographic one. You sent the pictures, so I allow myself this slip.

1. Clinical diagnosis: scleroderma. With this facies, without looking at the picture of the hands, this seems to me to the first diagnosis that comes to mind.

2. ECG; regular rhythm, 75 per minute, I cannot visualize P waves in DI and V5 seems to inscribe with PR of 0.60 sec. Shifted axis to the left by left anterior fascicular block. High R waves in V2 and V3, probably due to septal hypertrophy. Signs of LV overload. In scleroderma a musculo-cutaneous disorder may be found, with progressive proximal weakness and cardiac involvement, with development of hypertrophic cadiomyopathy. Regards,

Martin Ibarrola

Bloqueo aurículo-ventricular completo como signo de afección cardíaca en paciente con esclerodermia omero Requena, JM; Bureo Dacal, JC

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Severe cardiac involvement in children with systemic sclerosis and myositis. Pierre Quartier

http://www.jrheum.org/content/29/8/1767.short

Amigos: en ECG se ve onda P solo en V1 y V2. El ritmo pienso que es ectópico.
Presenta una hipertrofia de VI con hemibloqueo anterosuperior izquierdo, FC normal y ritmo regular, una extra-sístole ventricular. No se observa primer vector de QRS y en V1 parece un area inactiva.
QS en aVR (derivaciones derechas) pero podria ser debido a una miocardiopatia hipertrofica septal asimétrica donde el septum no se manifiesta debido a la anarquia de la arquitectura de las fibras musculares.
El aspecto de la paciente es el de una enfermedad sistémica quizas fibro muscular.
Saludos

Emilio Marigliano emiliomarigliano2000@yahoo.com

Ectopic regular rhythm with normal heart rate. I observe P wave only in V1-V2.
Isolated Premature ventricular contraction
Left Anterior Fascicular Block
Inactive electrical area in V1. QS patterm in right percordial lead aVR: Absence of the first vector? Consequence of asymmetrical hypertrophic cardiomyopathy?
The patient general aspect is suggestive of systemic disease probably fibro-muscular.
Greeting

FINAL COMMENTARIES

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

Clinical diagnosis: Scleroderma, progressive systemic sclerosis (PSS) or generalized scleroderma/CREST Syndrome. **PPS** is a systemic autoimmune disease of connective tissue in which there is thickening of dermal collagen bundles, and fibrosis and vascular abnormalities in internal organs.

PSS included:

1. Proximal cutaneous fibrosis
2. Raynaud phenomenon: present in 90% of cases and severe. Raynaud's normally affects the fingers and toes. Systemic scleroderma and Raynaud's can cause painful ulcers on the fingers or toes which are known as digital ulcers. Calcinosis (deposition of calcium in lumps under the skin) is also common in systemic scleroderma, and is often seen near the elbows, knees or other joints.
3. Gastrointestinal dysmotility
4. Pulmonary fibrosis with pulmonary hypertension. It may be due to intrinsic pulmonary artery disease or be secondary to interstitial fibrosis. Some impairment in lung function is almost universally seen in patients with diffuse scleroderma on pulmonary function testing;7 however, it does not necessarily cause symptoms, such as shortness of breath. Some patients can develop pulmonary hypertension, or elevation in the pressures of the pulmonary arteries. This can be progressive, and lead to right sided heart failure. The earliest manifestation of this may be a decreased diffusion capacity on pulmonary function testing.Other pulmonary complications in more advanced disease include aspiration pneumonia, pulmonary hemorrhage and pneumothorax
5. Cardiac involvement: pericardial involvement, fibrinous pericarditis present in 70% of cases at autopsy, eventual singnificative effusion. The presence of moderate to large effusion is an independent risk factor of mortality, Pthychy myocardial fibrosis, occasionally with contraction band necrosis, microvascular occlusion due to vasospasm, fixed perfusion defect on Thallium imaging observed in 95% of cases, myocardial infarction with normal coronaries vessels, ventricular conduction defects, septal pseudoinfact pattern, PVCs(>60% of cases), LV failure. Eventual syncope and SCD tendency.

6. Renal crisis associated with hypertension and rise of creatinine levels. Renal involvement, in scleroderma, is considered a poor prognostic factor and frequently a cause of death.The most important clinical complication of scleroderma involving the kidney is *scleroderma renal crisis*. Symptoms of scleroderma renal crisis are malignant hypertension (high blood pressure with evidence of acute organ damage), hyperreninemia (high renin levels), azotemia (kidney failure with accumulation of waste products in the blood) and microangiopathic hemolytic anemia (destruction of red blood cells). Apart from the high blood pressure, hematuria (blood in the urine) and proteinuria (protein loss in the urine) may be indicative.In the past scleroderma renal crisis was almost uniformily fatal. While outcomes have improved significantly with the use of ACE inhibitors the prognosis is often guarded, as a significant number of patients arerefractory to treatment and develop renal failure. Approximately 5-10% of all scleroderma patients develop renal crisis at some point in the course of their disease. Patients that have rapid skin involvement have the highest risk of renal complications. It is most common in diffuse cutaneous scleroderma, and is often associated with antibodies against RNA polymerase (in 59% of cases). Many proceed to dialysis, although this can be stopped within three years in about a third of cases. Higher age and (paradoxically) a lower blood pressure at presentation make it more likely that dialysis is needed.Treatments for scleroderma renal crisis include ACE inhibitors, which are also used for prophylaxis, and renal transplantation. Transplanted kidneys are known to be affected by scleroderma and patients with early onset renal disease (within one year of the scleroderma diagnosis) are thought to have the highest risk for recurrence

The limited CREST Syndrome (Calcinosis, Raynaud phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia) variant includes:

1. Calcinosis
2. Raynaud phenomenon
3. Eshophageal dysmotility
4. Sclerodactyly
5. Pulmonary hypertension
6. Fixed perfusion defect on Talium imaging observed in 65% of cases
7. Telangectasy

History: In 1910, Thibierge and Weissenbach described the first case report of what was later called CRST (calcinosis cutis, Raynaud phenomenon, sclerodactyly, and telangiectasia) syndrome in English by Winterbauer who, in 1964, described a series of 8 patients with the features that make up the abbreviation CRST.(1, 2) Although he noted esophageal dysmotility in 4 of 8 patients, he did not include this feature in his original description of CRST syndrome. Frayha et al(3) noted the frequent occurrence of esophageal dysmotility and suggested that the acronym CREST may be more appropriate. Velayos et al(4) reviewed 13 patients with CREST and CRST syndromes and found the syndromes equivalent.

1. **Meyer O. [From Thibierge-Weissenbach syndrome (1910) to anti-centromere antibodies (1980). Clinical and biological features of scleroderma]. *Ann Med Interne (Paris)*. Jan 1999;150:47-52.**
2. **Winterbauer RH. Multiple telangiectasia, Raynaud's phenomenon, sclerodactyly, and subcutaneous calcinosis: A syndrome mimicking hereditary hemorrhagic telangiectasia. *Bull Johns Hopkins Hosp*. Jun 1964;114:361-383.**
3. **Frayha RA, Scarola JA, Shulman LE. Calcinosis in scleroderma: A reevaluation of the CRST syndrome, abstracted. *Arthritis Rheum*. 1973;16:542.**
4. **Velayos EE, Masi AT, Stevens MB, Shulman LE. The 'CREST' syndrome. Comparison with systemic sclerosis (scleroderma). *Arch Intern Med*. Nov 1979;139:1240-1244.**

In 2004, Nadashkevich et al(1) proposed the classification criteria:

(1) Autoantibodies to centromere proteins, scl-70 (topo I) and fibrillarin;

(2) Bibasilar pulmonary fibrosis;

(3) Contractures of the digital joints or the prayer sign;

(4) Dermal thickening proximal to the wrists;

(5) Calcinosis cutis;

(6) Raynaud phenomenon (at least a 2-phase color change);

(7) Esophageal distal hypomotility or reflux esophagitis;

(8) Sclerodactyly or nonpitting digital edema; and

(9) Telangiectasias, which can be remembered by the abbreviation ABCDCREST.

Fulfilling 3 or more criteria indicates definite systemic scleroderma with a sensitivity and specificity as high as 99% and 100%, respectively.

1. **Maricq HR, Valter I. A working classification of scleroderma spectrum disorders: a proposal and the results of testing on a sample of patients. *Clin Exp Rheumatol*. Jan-Feb 2004;22(3 Suppl 33):S5-13.**

Also in 2004, Maricq and Valter(13) had a complex but potentially very useful proposal for classifying the scleroderma spectrum disorders; however, in 2005, Wollheim(14) reported that without substantial independent confirmatory work, this classification system may not gain widespread acceptance in its present form.
The Maricq and Valter(13) proposed classification for scleroderma spectrum disease is as follows:

Type I - Diffuse skin involvement proximal to elbows/knees; includes trunk
Type II - Intermediate skin involvement proximal to the metacarpal phalangeal/metatarsal phalangeal joints, distal to the elbows/knees; trunk not involved
Type III - Digital sclerodactyly only (meets American College of Rheumatology minor criteria but excludes those without skin involvement)
Type IV - Scleroderma sine scleroderma (capillary pattern or pitting scars and visceral involvement; no anticentromere antibodies; no telangiectasia)
Type V - Undifferentiated connective-tissue disease with 2 of 3 of the following scleroderma features: sclerodactyly, pitting scars, or scleroderma capillary pattern; or one of these features along with one of the following: Raynaud phenomenon, pulmonary fibrosis, or visceral involvement (esophagus, heart, kidney); but do not meet the criteria for groups III and IV; no anticentromere antibodies; no telangiectasia
Type VI - CREST; no skin involvement, or sclerodactyly only, telangiectasia is required with one or more other acronyms; or anticentromere antibodies are required with any 2 or more acronyms.

1. **Maricq HR, Valter I. A working classification of scleroderma spectrum disorders: a proposal and the results of testing on a sample of patients. *Clin Exp Rheumatol*. Jan-Feb 2004;22(3 Suppl 33):S5-13.**
2. **Wollheim FA. Classification of systemic sclerosis. Visions and reality. *Rheumatology (Oxford)*. Oct 2005;44:1212-1216.**

Scleroderma in a 1680 painting

The painting “Archangel Raphael with Bishop Domonte” by Murillo (1618-1682) is on display in the Pushkin Museum of Fine Arts in Moscow.

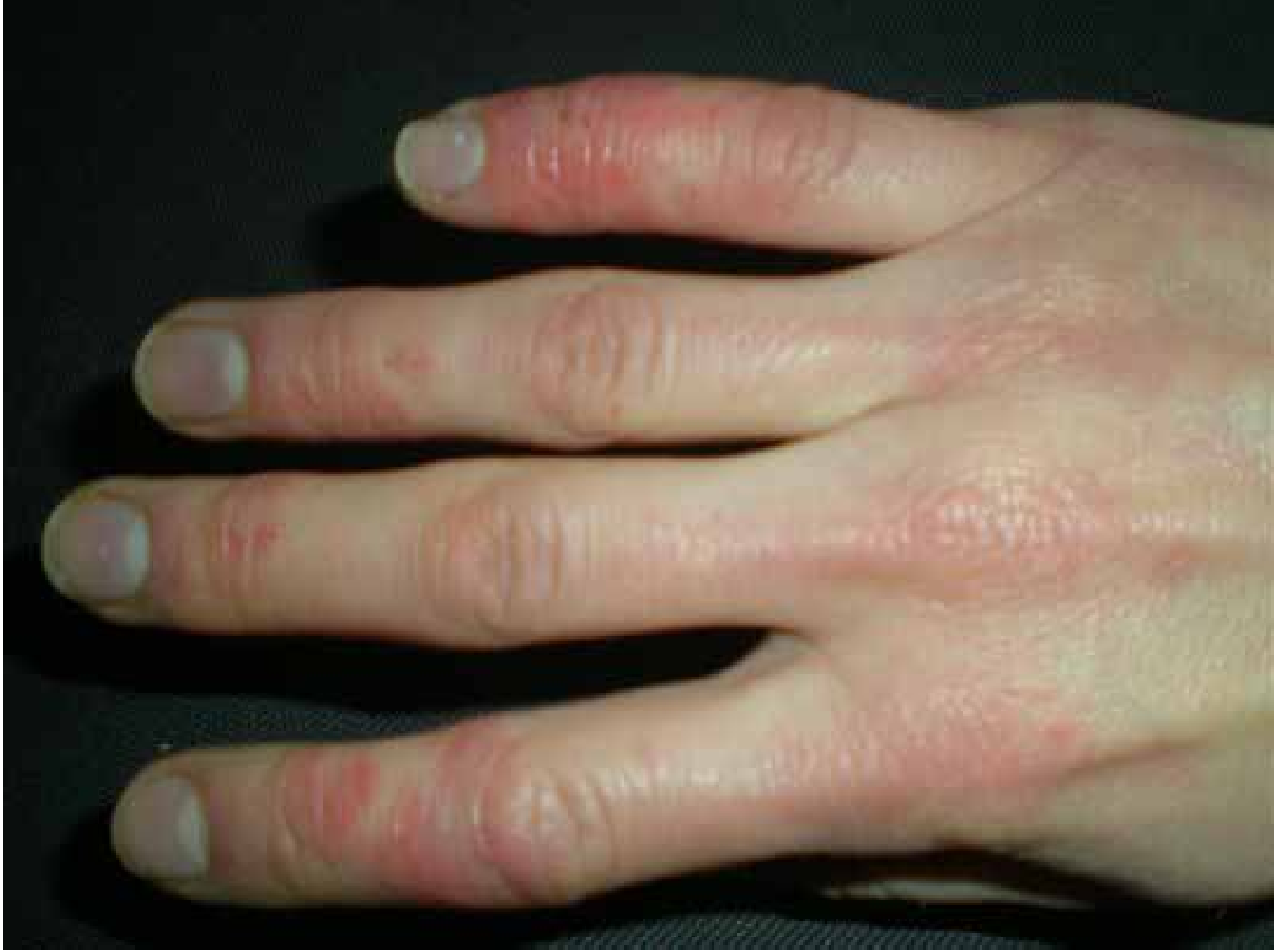
The portrait of Bishop Francisco Domonte, in the right bottom corner of the painting, shows several characteristics of scleroderma, with the most outstanding being the numerous telangiectasias in the face, lips and hands, as well as the tension of the skin around the nose, in the cheeks and the forehead. In the hands of the bishop, the skin folds over the joints are not visible and the fingers seem globally swollen. The term scleroderma refers to a characteristic fibrotic hardening of the skin.

Systemic sclerosis (the systemic form of scleroderma) is a rare disease, that affects both the skin and internal organs, which originates from superproduction and availability of collagen, basically being characterized by fibrosis of the structures involved.

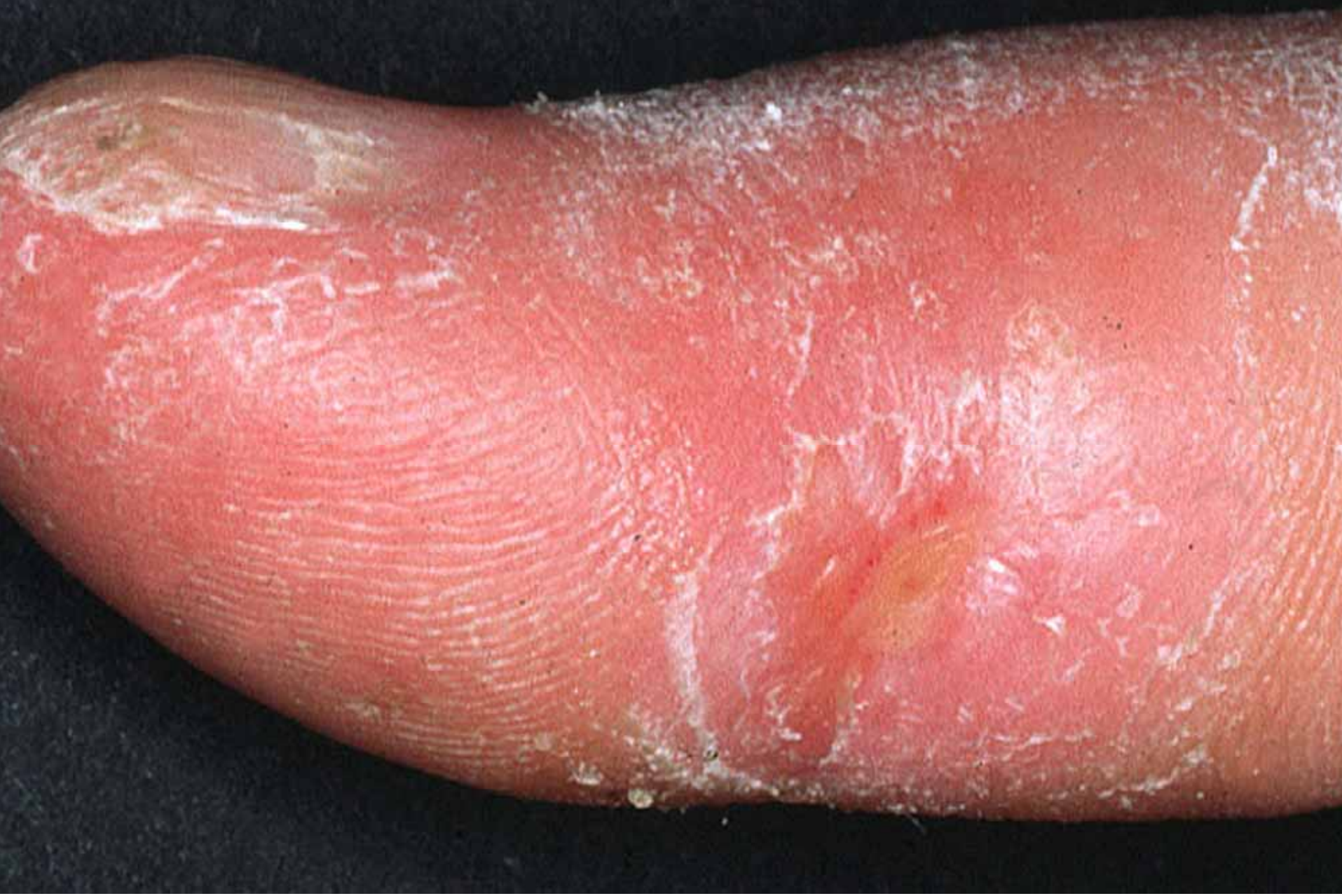
Skin manifestations are a framework of systemic sclerosis.

*Perhaps nothing is so eloquent and significant as the human face. Looking at one another is our most basic form of conversation, and wherever people meet in a primary or face-to-face relationship it is the face, which is generally the center of attention. It is the source of vocal communication, the expressor of emotions, and the revealer of personality traits. The face is the person him/herself. (Macgregor, 1951, p. 630).**I**f, as the above author contends, the human face is deeply and profoundly significant, what happens when an individual "loses face," both literally and figuratively? What happens when an individual, as the result of an illness, accident, or injury, is confronted with an unwanted and unanticipated change in facial appearance? What happens when there is very little, if anything, an individual can do to remedy this change? These are certainly some of the questions, even if unspoken, that many individuals with scleroderma-related facial changes live with on a daily basis as they move through their personal and social worlds, catching a glimpse of themselves in a mirror or store window, or in the reaction of others around them, be they significant or mere strangers. These are questions that may never have a satisfactory resolution; yet they are questions whose answers will ultimately define the individual who has been asked.*

Typical Scleroderma-Related Facial Changes



The fingers have characteristic alterations: they are swollen and the skin becomes thicker, firmly linked to the underlying subcutanous tissue. The normal skin folds over the joints disappear gradually. These fibrotic alterations in the fingers are known as sclerodactyly, that although in isolation are not pathognomonic to scleroderma, when the fibrotic alterations are near the metacarpophalangeal joints (i.e. reach the back of the hands), the diagnosis of scleroderma is virtually defined.



Clinical appearence of acrosclerotic piece-meal necrosis of the first digit in a patient with systemic sclerosis.

Systemic Scleroderma-Related Facial Changes

As is true for scleroderma in general, facial changes vary in their course and effects from patient to patient and over time in any given individual. In the majority of cases, however, facial appearance for individuals with systemic and linear scleroderma *en coup de sabre* is altered slowly over time, with changes occurring over several months to a number of years.

In systemic scleroderma there are three recognized phases of skin thickening during the facial change process: the **edematous**, **indurative**, and **atrophic** phases (Clements & Medsger, 1996). These phases generally follow one another as the illness progresses.

In the **edematous** phase, pitting or nonpitting edema (swelling) may occur in the face and last indefinitely, but the swelling is usually painless.

During the **indurative** phase, the edema is gradually replaced by a markedly thickened dermis and an increasingly thinned epidermal layer, leading to the loss of skin creases and a "choking out" of hair follicles, sweat glands, and sebaceous glands. The latter process is caused by collagenous deposition in the upper dermis.

In the diffuse form of scleroderma, this process occurs over several months or years, whereas in the limited form of scleroderma (known as the CREST syndrome), this phase may occur slowly over several years, and up to 20 years.

In terms of facial changes, this entire process results in a "characteristic 'pinched nose' or 'mauskopf' appearance, sometimes interpreted as an 'expressionless' facies due to reduced mobility of the eyelids, cheeks, nose, and mouth during ordinary conversation" (Clements & Medsger, 1996, p. 390).

In the **atrophic** phase, usually occurring after several years, the thickened dermis softens and either reverts to normal thickness or actually becomes thinner than normal.

A "binding down" or "tethering" occurs when the dermis becomes more firmly attached to underlying subcutaneous fat.

In this late stage, telangiectasias (clusters of dilated and tortuous capillaries and venules) increase in number and tend to be the dominant visual feature, appearing as a single red spot or in clusters on both cheeks and over the nose (Clements & Medsger, 1996). The latter shape resembles a butterfly and is known as the "butterfly-look."
The vast majority of functional problems that result from systemic scleroderma-related facial changes are due to the effects of microstomia or "small mouth."
In addition to physical pain, the effects of microstomia include:

- Rightly pursed lips that lose their fullness;
- Peri oral subcutaneous fibrosis; and
- Temporomandibular joint involvement.

These effects all contribute to reduced oral aperture and radial furrowing around the mouth.
Lip retraction, which accentuates the prominence of the central incisor teeth, also makes normal mouth closure impossible (Clements & Medsger, 1996).
Microstomia greatly interferes with several critical functions including eating, speaking, and proper prophylactic oral hygiene.

Many scleroderma patients also suffer from a constriction in the movement of the tongue.
If present, secondary Sjögren's (sicca) syndrome, which interferes with the normal production of saliva, may also offer an additional threat to dentition, that is, the appearance and arrangement of the teeth (Clements & Medsger, 1996).

"Secondary" Sjögren's syndrome, as opposed to "primary," refers to the two forms of this disorder. Secondary Sjogren's syndrome occurs with rheumatic diseases such as rheumatoid arthritis, systemic lupus erythematosus and scleroderma.

Primary Sjögren's syndrome occurs by itself and is not associated with other diseases.
Sjögren's syndrome is an autoimmune disease in which lymphocytes invade and damage moisture-producing glands, primarily of the eyes and mouth, preventing them from producing tears and saliva, respectively (Arthritis Foundation, 1997).
In addition to the physical and functional problems associated with microstomia, there are social and relational side-effects. For example, eating and speaking generally imply human interaction; when these functions are compromised, so too is one's interactional ease and capability.
The effects of microstomia may also limit or interfere with acts of affection and sexuality—vital aspects of human relationships.
In essence, this particular symptom has the capacity to compromise an individual on many different levels – physically and emotionally; personally and interpersonally.
A cohort of 405 patients with Systemic sclerosis SSc was identified retrospectively by Hashimoto et al (1). Data on clinical features, including autoantibodies, organ involvement, and overlap of other connective tissue diseases, were studied. The percentage of male patients during or after 1990 was greater than that before 1990 (3.9 vs. 10.6%, respectively).
Limited cutaneous SSc (lSSc) was twice as frequent as diffuse cutaneous SSc (dSSc). About half of the patients had lung involvement (50.4%), while only 3.2% had scleroderma renal crisis. Male gender was associated with lung involvement, and dSSc was associated with most organ involvements except for pulmonary arterial hypertension (PAH). Anti-Scl-70 antibody was associated with lung or heart involvement, while anti-U1-RNP antibody was only associated with PAH. Patients with anti-centromere antibody had less organ involvement. SSc-Sjögren overlap syndrome was related to lSSc, further overlapping systemic lupus erythematosus (SLE), and less lung or heart involvement. The authors observed an increased proportion of male patients in recent years.

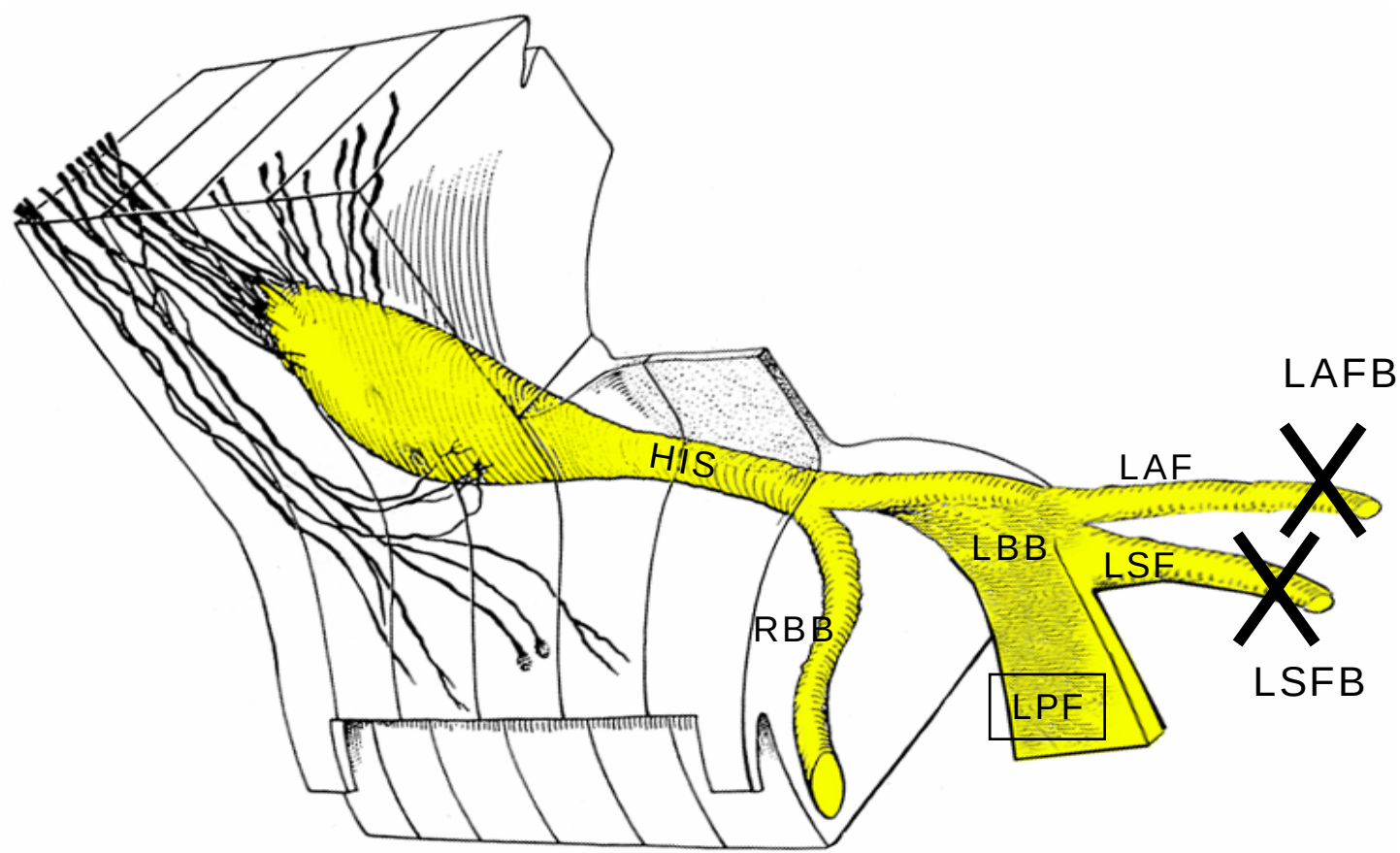
1. **Hashimoto A, Endo H, Kondo H, Hirohata S. Clinical features of 405 Japanese patients with systemic sclerosis.Mod Rheumatol. 2011 Aug 28. [Epub ahead of print]**

Clinical and epidemiological findings indicate that symptomatic heart disease in patients with systemic sclerosis predicts poor prognosis, but cardiac involvement may occur years before clinical manifestation. Low heart rate variability, and the presence of anti-SCL70 are correlated with preclinical cardiac involvement in SSc patients and may predict the likelihood of malignant arrhythmia and sudden cardiac death. Therefore, noninvasive HRV evaluation before clinical cardiac involvement in these patients might be beneficial when added to the clinical and laboratory assessments in detecting high-risk patients, and may allow for implementation of preventive measures and initiation of appropriate therapy early in the course of the disease.
Holter analysis for Systemic Scleroderma patients revealed an increased prevalence of PVCs ≥ 10/h and supra-ventricular tachycardias. Total skin thickness score, Raynaud's phenomenon and anti-scleroderma 70 (anti-SCL70) show significant positive correlations with all arrhythmia parameters, while showing a significant negative correlation with the impaired ventricular diastolic function and various HRV parameters. No correlation was found between arrhythmia and HRV parameters and disease duration, disease type, or presence of anti-centromere antibodies.(1)
At a preclinical level, is important in all the cases as an asymptomatic patient may have diastolic dysfunction which can be treated and should be closely observed.(2) The evaluation of LV diastolic function is an essential component of the echocardiographic examination for dyspneic patients with impaired or preserved LV systolic function. Doppler echocardiography in combination with two-dimensional echocardiographic findings can assist the diagnosis of underlying cardiac dysfunction, give an estimate of LV filling pressures, guide heart failure treatment, and provide important prognostic information.

1. **Othman KM, Assaf NY, Farouk HM, Aly Hassan IM. Autonomic dysfunction predicts early cardiac affection in patients with systemic sclerosis.Clin Med Insights Arthritis Musculoskelet Disord. 2010 May 24;3:43-54.**
2. **Poanta L, Dadu R, Tiboc C, Rednic S, Dumitrascu D. Systolic and diastolic function in patients with systemic sclerosis. Eur J Intern Med. 2009 Jul;20:378-82.**

ECG diagnosis

Rhythm: Atrial fibrillation QRS axis: extreme left axis deviation on frontal plane: -45º, SIII>SII, final s waves in left precordial lead: Left Anterior Fascicular Block (LABF)
QRS duration 105ms, Qr in V1, qRs from V2 to V4, Prominent Anterior Forces (PAF)
Conclusions:
Atrial fibrillation? We don't observe P wave but QRS complexes are regular or near regular
Isolated premature ventricular contraction
Left bifascicular block: LAFB + Left Septal Fascicular Block (LSFB)? RVH? Dorsal infarction?



ECG-VCG CRITERIA FOR LSFB

A) ELECTROCARDIOGRAPHIC:

1) Normal QRS duration or with a discrete increase (up to 110 ms).
2) FP leads with no modifications: normal QRS.
3) Increased intrinsicoid deflection of V_1 and V_2.
4) R wave voltage of $V_1 \geq$ than 5 mm;
5) R/S ratio in V_1 > 2;
6) R/S ratio in V_2 > 2;
7) S wave depth in V2 < 5 mm.
8) Possible small q wave in V2 or V1 and V2.
9) R wave of V2 > 15 mm;
10) RS or Rs in V2 and V3 (frequent rS in V1) with R wave "in crescendo" from V1 to V2 or V3 and decreasing from V5 to V6;
11) Absence of q wave in V5, V6 and DI (by absence of the vector 1AM). First 10ms initial QRS vector.

Electrocardiographic criteria of the LSFB.
In red color the criteria present in this case

1. **Pérez Riera AR, Ferreira C, Ferreira Filho C, Meneghini A, Uchida AH, Moffa PJ, Schapachnik E, Dubner S, Baranchuk A. Electrovectorcardiographic diagnosis of left septal fascicular block: anatomic and clinical considerations. Ann Noninvasive Electrocardiol. 2011 Apr;16(2):196-207..**

ECG-VCG CRITERIA FOR LSFB

B) VECTOCARDIOGRAPHIC: (all in the HP)

1) QRS loop in the HP with an area predominantly located in the left anterior quadrant (> 2/3 of the loop facing the orthogonal X line);
2) Absence of normal convexity to the right of the initial 20 ms of the QRS loop;
3) Discrete dextro-orientation with moderate delay of the vector from 20 ms to 30 ms;
4) Anterior location of the vector from 40 to 50 ms;
5) Posterior location with a reduced magnitude of the vector from 60 to 70 ms;
6) Maximal vector of the QRS loop located to the right of + 30º;
7) T loop of posterior orientation (useful for the differential diagnosis with basal inferior or dorsal infarction);
8) The QRS loop rotation may be:
   - (8a) Counterclockwise: incomplete LSFB.
   - (8b) Clockwise: advanced LSFB or in association with CRBBB, LAFB or LPFB.

Vectorcardiographic criteria of the LSFB.

Commentaries: The Brazilian Guidelines for Interpreting Rest Electrocardiogram (*1*) provided us with the following criteria for ECG diagnosis of LSFB:

QRS duration < 120 ms, in general, close to 100 ms. The appearance of LFB does not increase QRSD by more than 25 ms, due to multiple interconnections between the fascicles of the LBB ("passage way zone" of Rosenbaum). The QRS complex is slightly prolonged between 100 ms to 115 ms. Thus, LSFB pattern with a prolonged QRSD indicates the presence of additional conduction disturbances such as: other fascicular blocks, RBBB, MI, focal block, or a combination of these;

15 mm R waves in V2 and V3 or from V1;

Increasing for all intermediary precordial leads and decreasing from V5 to V6;

"r" wave jump may occur from V1 to V2 ("rS" in V1 for R in V2);

Absence of initial q wave in left leads

Absence of SÂQRS shift if isolated. In this case, extreme left axis deviation because it is associated with LAFB: Left bifascicular block

T wave most of the times, negative in right precordial leads.

Observation: all these criteria are valid in absence of RVH, septal hypertrophy or dorsal or basal inferior MI/fibrosis and other causes of Prominent Anterior Forces (PAF).

In the present case we have the possibility that PAF be secondary to RVH consequence of Pulmonary fibrosis with pulmonary hypertension. It may be due to intrinsic pulmonary artery disease or be secondary to interstitial fibrosis. Additionally we have the possibility of intraventricular conduction defect: LAFB associated with LSFB. Another possibility is the presence of basal fibrosis(old dorsal infarction) and PAF.

1. **Pastore CA et al. Guidelines for Interpreting Rest Electrocardiogram. Arq Bras Cardiol; 2003; 80: 1-17 Arq Bras Cardiol; 2003; 80: 1-17.**

PROMINENT ANTERIOR FORCES(PAF) DEFINITION BY ELECTROCARDIOGRAPHIC PARAMETERS

We consider there is presence of PAF in ECG, when the voltage of R wave in any precordial lead of the anterior or anteroseptal wall from V1 (+115º) through V4 (+47º) is greater than the normal maximal limit for gender and age. Electro-vectocardiographic criteria of PAF should be age-related and gender-related. Thus, in lead V1 in adults between 20 and 30 years old, R wave > 8.9 mm in women and in men > 5.3 mm is considered a criterion for PAF.

From 30 to 40 years old, in women R wave voltage > 5.4 mm and in men > 5.8 mm is considered a criterion for PAF. Finally, from 40 to 60 years old, R wave > 4.9 mm in women, and > 4.0 mm in men is considered a criterion for PAF. (*1-4*).

Another criterion used by some authors to consider the presence of PAF regards the R/S ratio in V1. Thus, an R/S ratio in V1 ≥1 is considered abnormal in adults. Tall lead V1 (tall R V1) is defined as an R/S ratio equal to or greater than 1. From our point of view, this criterion with these values cannot be considered as valid, since in 1% of normal individuals this ratio (R/S ratio in V1 ≥1) is found as a normal variant.

In lead V2, approximately in 25% of men and 12% of women the R/S ratio is 1.

From 20 to 30 years old, R wave > 13.9 mm in women and > 9.2 mm in men is considered a criterion for PAF.

From 30 to 40 years old, R wave > 12.1 mm in women and > 10.1 mm in men is considered a criterion for PAF.

From 40 to 60 years old, R wave > 12.0 mm in women and > 9.1 mm in men is considered a criterion for PAF. This woman has 27mm of R wave in V2

1. **Macfarlane PW, Lawrie TDV, eds. The normal electrocardiogram and vectorcardiogram. In: Comprehensive Electrocardiology: Theory and Practice in Health and Disease. Vol 1-3. New York, NY: Pergamon Press, 1989.**
2. **Mattu A, Brady WJ, Perron AD, et al. Prominent R wave in lead V1: electrocardiographic differential diagnosis. Am J Emerg Med. 2001; 19: 504-513.**
3. **Yang TF, Macfarlane PW. Normal limits of the derived vectorcardiogram in Caucasians. Clin Physiol. 1994; 14: 633-646.**
4. **Yang TF, Chen CY, Chiang BN, Macfarlane PW. Normal limits of derived vectorcardiogram in Chinese. Electrocardiol. 1993; 26: 97-106.**

The normal amplitudes of R waves in lead V3 are:From 20 to 30 years old, R wave > 11.6 mm in women and > 8.2 mm in men is considered a criterion for PAF.

From 30 to 40 years old, R wave > 9.4 mm in women and > 7.1 mm in men is considered a criterion for PAF.

From 40 to 60 years old, R wave > 8.4 mm in women and > 7.1 mm in men is considered a criterion for PAF.

Finally, the normal amplitudes of R waves in lead V4 are:

From 20 to 30 years old, R wave > 27.7 mm in women and > 19.6 mm in men is considered a criterion for PAF.

From 30 to 40 years old, R wave > 29.2 mm in women and > 25.9 mm in men is considered a criterion for PAF.

From 40 to 60 years old, R wave > 25.6 mm in women and > 23.6 mm in men is considered a criterion for PAF.

Possible causes of PAF Differential diagnosis

1. Normal variant: counterclockwise rotation in the longitudinal axis.
2. RVE types A and B.
3. LVE of the diastolic or volumetric type.
4. Strict or posterior dorsal infarction.(New nomenclature basal inferior MI
5. Postero-lateral or postero-latero-inferior infarction.
6. Right bundle branch block.
7. Wolff-Parkinson-White with anomalous bundle of posterior location: Type A
8. Hypertrophic cardiomyopathy: obstructive and non-obstructive form.
9. Duchenne-Erb disease, pseudo hypertrophic muscular dystrophy linked to sex or infantile malignant (DMD).
10. Left Septal Fascicular Block (LSFB).
11. Associations of the previous ones: E.g.: RVE + CRBBB (next slide)

Possible causes of LSFB

1. Chronic Chagas cardiomyopathy (**1;2**)
2. Coronary Artery Disease (CAD): critical lesion of LADA and/or its septal perforating branches before the first septal (S1) one(**3;4**)
3. Coronary Artery Disease with Wellens syndrome (**5**)
4. Diabetes Mellitus (**6**) ?
5. Non-Obstructive Hypertrophic Cardiomyopathy (NO-HCM) (7)
6. Obstructive Hypertrophic Cardiomyopathy (O-HCM)(7);Nambiar et al.(8) presented a 39 year old male with recurrent VT developed features of progressive systemic sclerosis associated with O-HCM. The condition was diagnosed during life with skin biopsy. Myocardial involvement was detected by 2-D echocardiography and was confirmed at autopsy.(8)
7. Papillary Muscle Dysfunction (9)?
8. Kearns-Sayre Syndrome (10).

1) **Vichi FL. Et al. The prevalence of branch and left fascicular blocks in the bundle of His in Chagas' cardiomyopathy. Arq Bras Cardiol 1982;39:87- 88.**
2) **Moffa PJ, et al. The left anterior septal block in Chagas' disease. Jap Heart J. 1982; 23:163-165.**
3) **Moffa PJ, et al. The left-middle (septal) fascicular block and coronary heart disease. In Liebman J, ed. Electrocardiology'96 –From the cell to body surface. Cleveland, Ohio, Word Scientific, 1996;547-550.**
4) **Uchida AH, et al .Exercise-induced left septal fascicular block: an expression of severe myocardial ischemia. Indian Pacing Electrophysiol J. 2006 Apr 1; 6: 135-138.**
5) **Riera AR, et al. Wellens syndrome associated with prominent anterior QRS forces: an expression of left septal fascicular block? J Electrocardiol. 2008 Nov-Dec; 41: 671-674.**
6) **Magnacca M, al. Diagnostic value of electrocardiogram in septal fascicular conduction disorders of the left branch in diabetics Minerva Cardioangiolica 1988; 36:361-363.**
7) **Cheng CH, et al, - ECG pattern of left ventricular hypertrophy in non obstrutive hypertrophic cardiomiopathy: The significance of the mid-precordial changes. Am Heart J 1979; 97: 687-695.**
8) **Nambiar CA, et al. Progressive systemic sclerosis presenting as recurrent ventricular tachycardia and hypertrophic cardiomyopathy--a case report. J Assoc Physicians India. 1989 Feb;37:179-81.**
9) **Kobashi A. et al. Solitary papillary muscle hypertrophy as a possible form of hypertrophic cardiomyopathy. Jpn Circ J. 1998 Nov; 62: 811-816.**
10) **Riera AR, et al. Kearns-Sayre syndrome: electro-vectorcardiographic evolution for left septal fascicular block of the his bundle J J Electrocardiol. 2008 Nov-Dec; 41: 675-678.**

Hypothetical model of the sequence of ventricular activation in LSFB associated to LAFB.

In 1970, Dr. Dirk Durren et al(1) from the University Department of Cardiology and Clinical Physiology, Wilhelmina-Gasthuis, Amsterdam, The Netherlands, demonstrated in a classical manuscript using 870 intramural terminals in isolated human hearts, that three endocardial areas are synchronously excited from 0 to 5 ms after the start of left ventricle (LV) activity potential (See Figure next slide). To obtain information concerning the time course and instantaneous distribution of the excitatory process of the normal human heart, the authors studied on isolated human hearts from seven individuals who died from various cerebral conditions, but who had no history of cardiac disease. The first LV areas excited were:

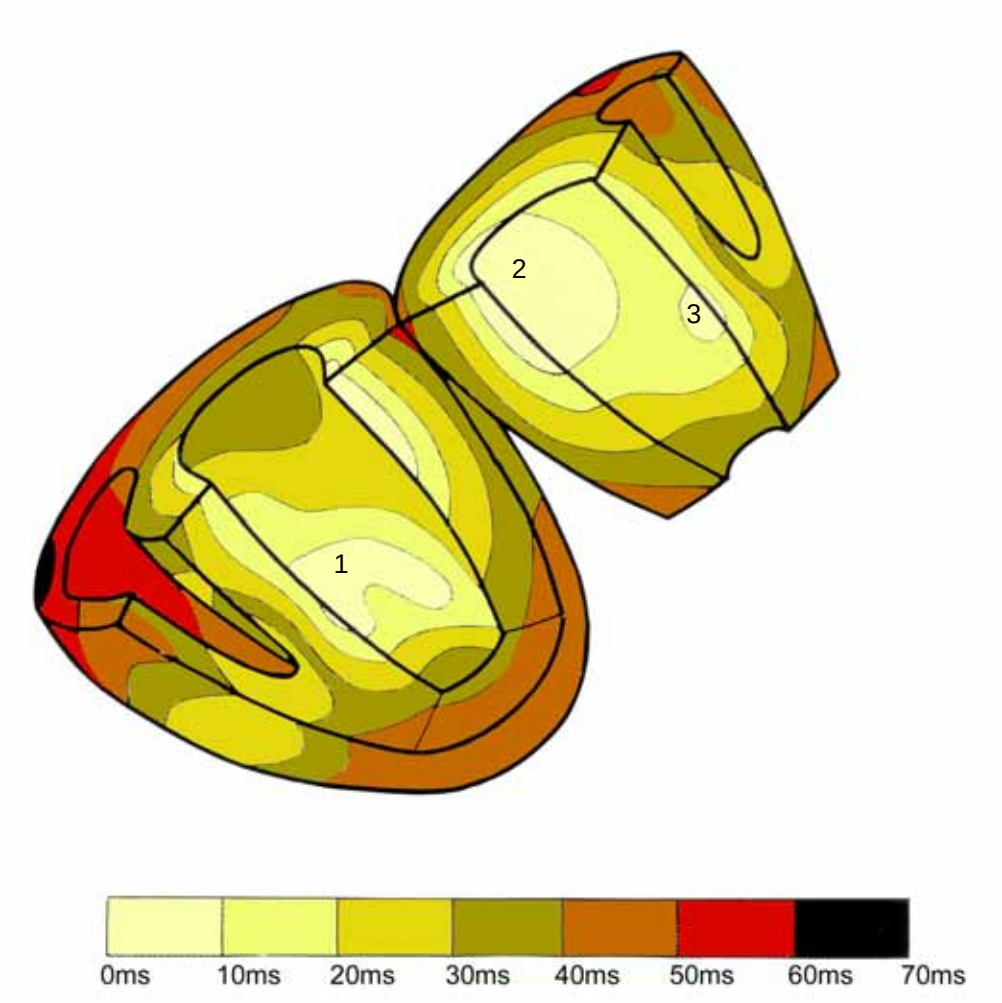
1. High on the anterior paraseptal wall just below the attachment of the ALPM where the LAF ends

2. Central on the left surface of the IVS where the LSF ends. Septal activation started in the middle third of the left side of the IVS, somewhat anteriorly and the lower third at the junction of the IVS and posterior wall. The normally functioning LSF, the left middle septum surface and the inferior two-thirds of the septum originate the first vector, vector 1 or first anteromedial (1AM) vector (*16*) and

3. Left inferior two-thirds of the IVS (second vector or vector of the inferior 2/3 of IVS) (*17*); Posterior paraseptal about one third of the distance from the apex to the base near the base of PMPM where the LPF ends. The posterobasal area is the last part of the LV to activate.

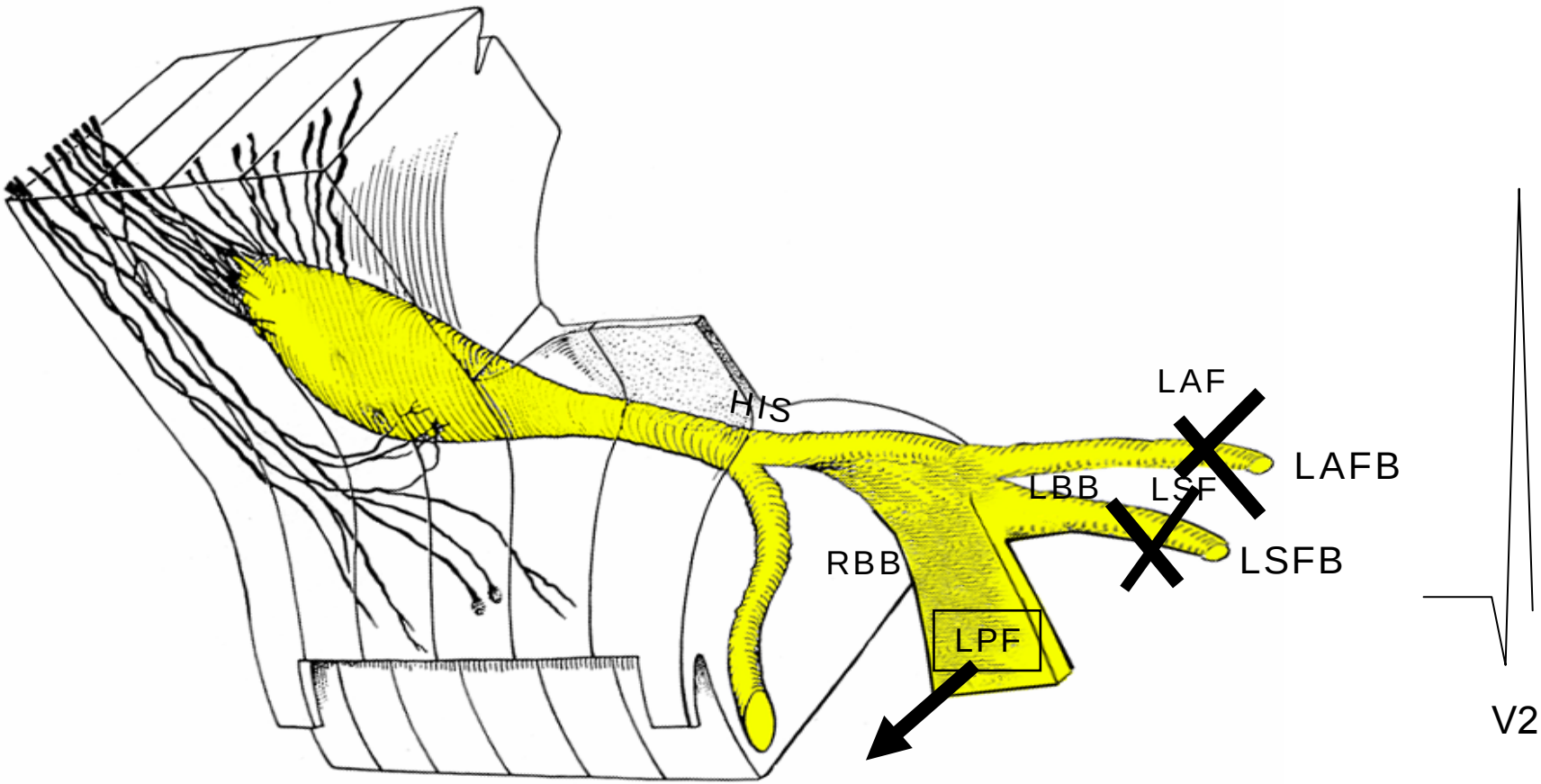
1. **Durrer D, van Dam RT, Freud GE, et al. Total excitation of the isolated human heart. Circulation 1970; 44: 899-912.**

INITIAL NORMAL QRS DEPOLARIZATION IN THREE POINTS



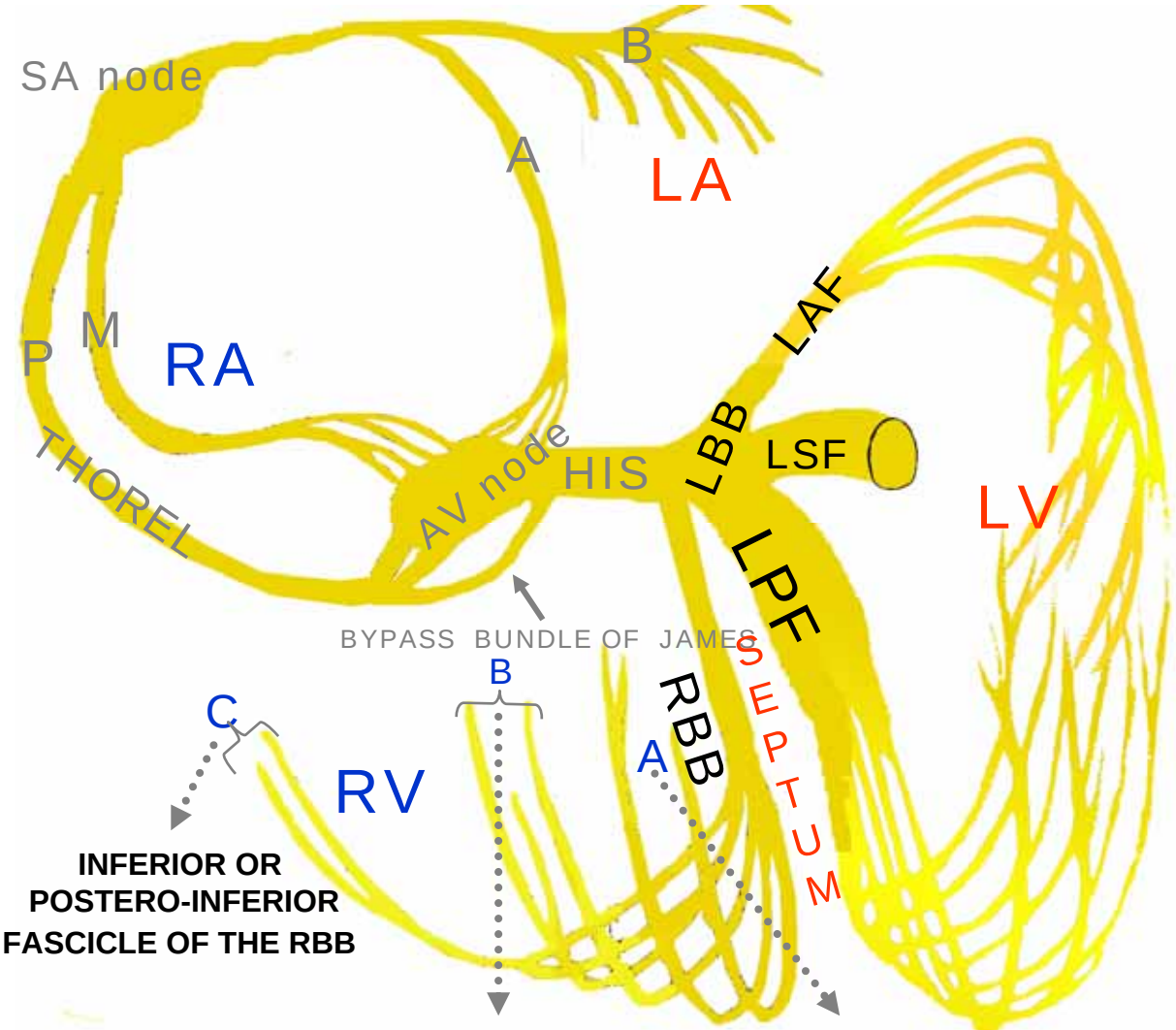
Initial 0ms to 10 ms
1) CENTRAL & APICAL REGION: LSF 2) ANTERO-SUPERIOR REGION: LAF 3) POSTERO-INFERIOR REGION: LPF

LAFB + LPFB INITIAL ACTIVATION



The first vector is dependent of LPF and directed to back: initial q wave in right precordial leads

COMPONENTS OF THE CARDIONECTOR SYSTEM OF SINO-ATRIOVENTRICULAR AND INTRAVENTRICULAR CONDUCTION



INTERNODAL BUNDLES

A **ANTERIOR**

M **MIDDLE OR OF WENCKBACH**

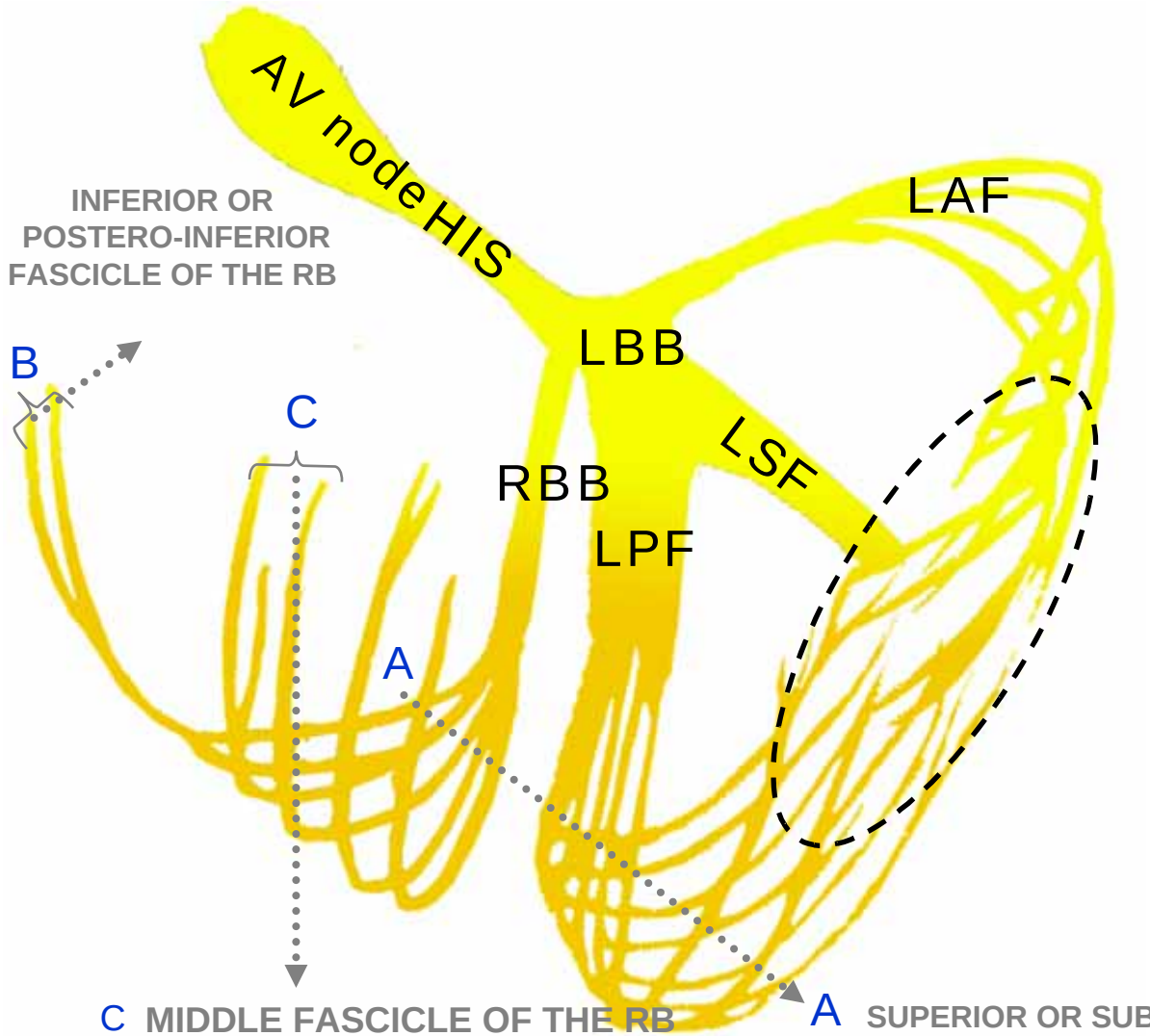
P **POSTERIOR OR OF THOREL**

B **BACHMAN'S FASCICLE (IT ACTIVATES THE LA)**

Affected in Brugada syndrome

Cardionector system of the heart: sino-atrioventricular and intraventricular.

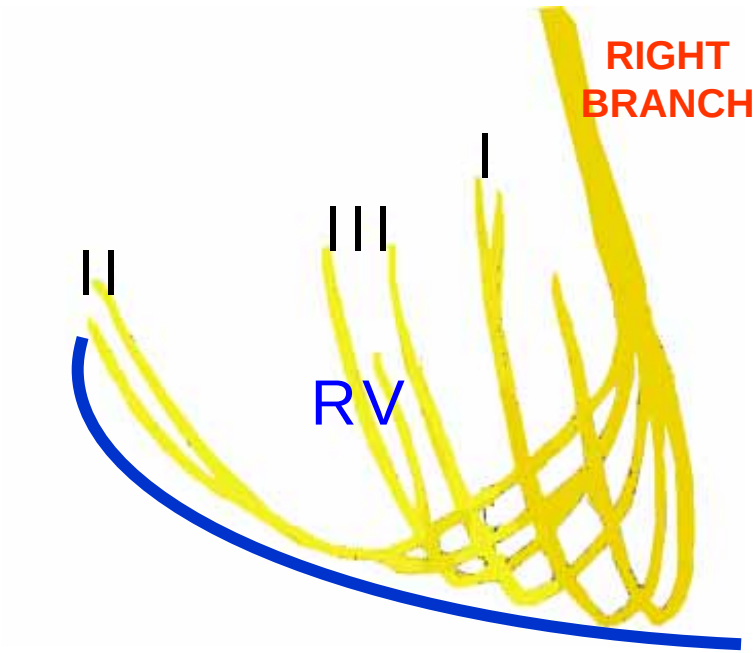
THE HEXAFASCICULAR NATURE OF THE HUMAN INTRAVENTRICULAR HIS SYSTEM

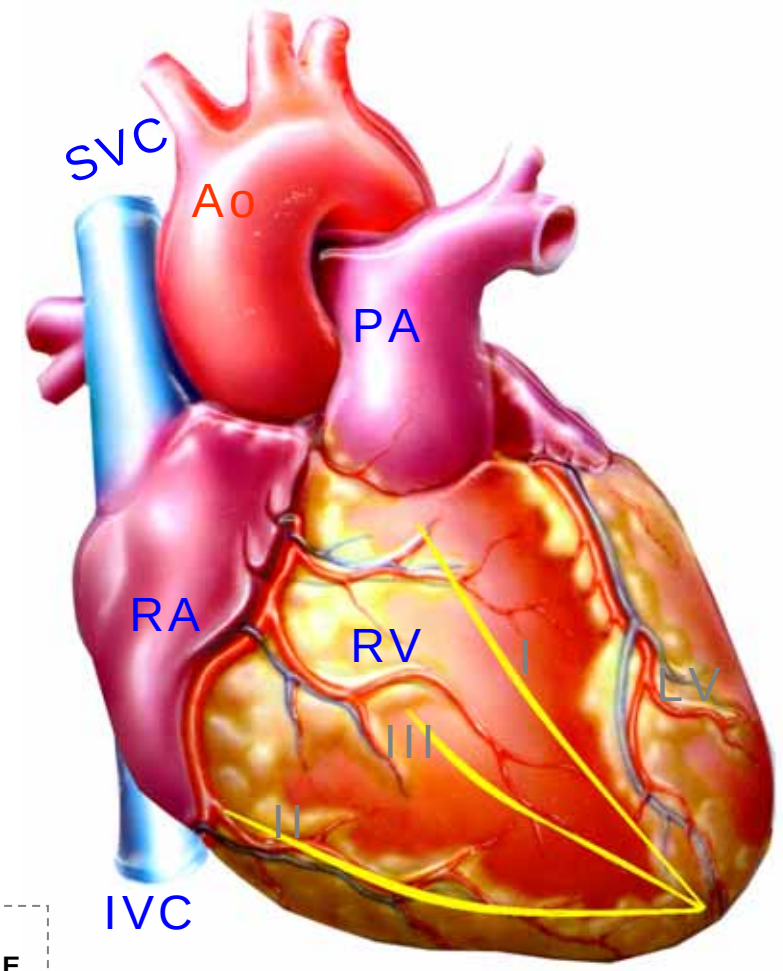


RBB: Right branch of the His Bundle.
LAF: Left anterior fascicle.
LSF: Left septal fascicle.
LPF: Left posterior fascicle

Our concept of hexafascicular nature of the human intraventricular His system.

DISTRIBUTION OF THE THREE FASCICLES OF THE RIGHT BRANCH OF THE HIS BUNDLE IN THE RV FREE WALL

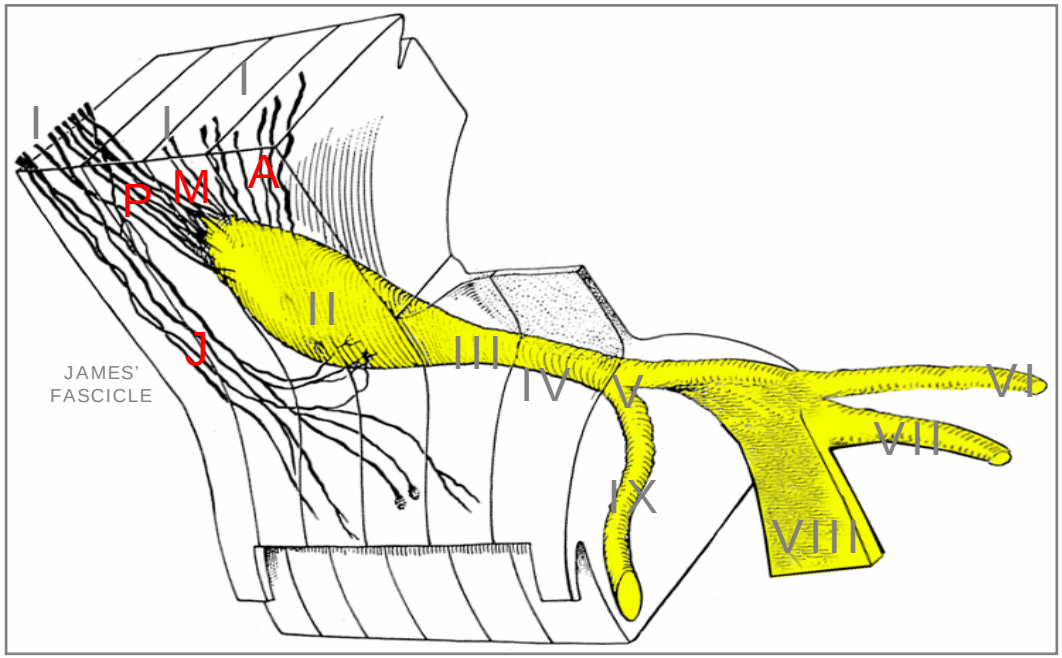


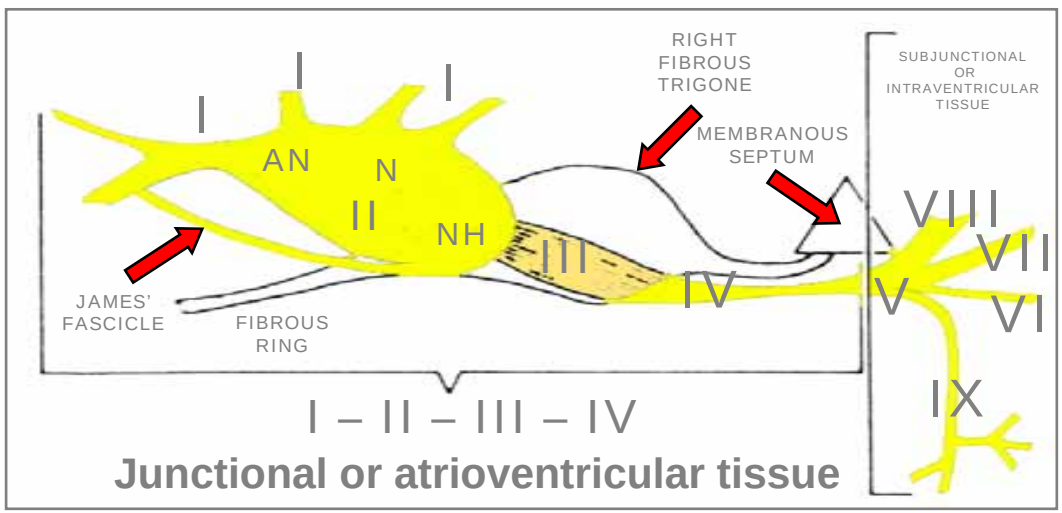


I - TERRITORY OF THE SUPERIOR OR SUBPULMONARY FASCICLE
II – TERRITORY OF THE INFERIOR OR POSTERO-INFERIOR FASCICLE
III – TERRITORY OF THE MIDDLE FASCICLE

Distribution of the three fascicles of the right branch in the RV free wall.

COMPONENTS OF THE ATRIOVENTRICULAR AND INTRAVENTRICULAR SYSTEM

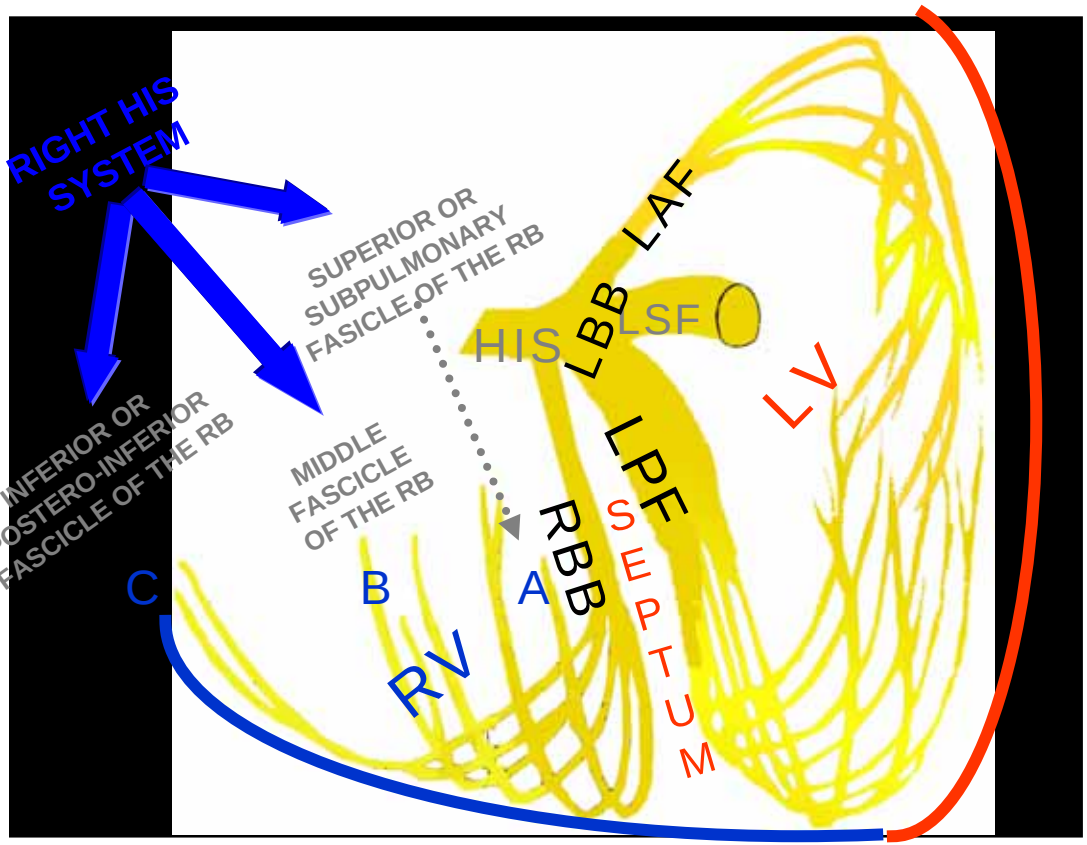




I	ANTERIOR, MIDDLE AND POSTERIOR INTERNODAL BUNDLES ENTERING INTO THE AV NODE
II	AV NODE
III	PENETRATING PORTION OF THE HIS BUNDLE
IV	NON BRANCHING NON PENETRATING PORTION OF THE HIS BUNDLE
V	BRANCHING PORTION OF THE HIS BUNDLE
VI	LAF
VII	LSF
VIII	LPF
IX	RB
I – II – III – IV	JUNCTIONAL TISSUE
V – VI – VII – VIII – IX	SUBJUNCTIONAL TISSUE

The atrioventricular or junctional and intraventricular or subjunctional system.

THE HEXAFASCICULAR NATURE OF THE HIS SYSTEM: THREE OF THE LEFT SYSTEM AND THREE OF THE RIGHT SYSTEM



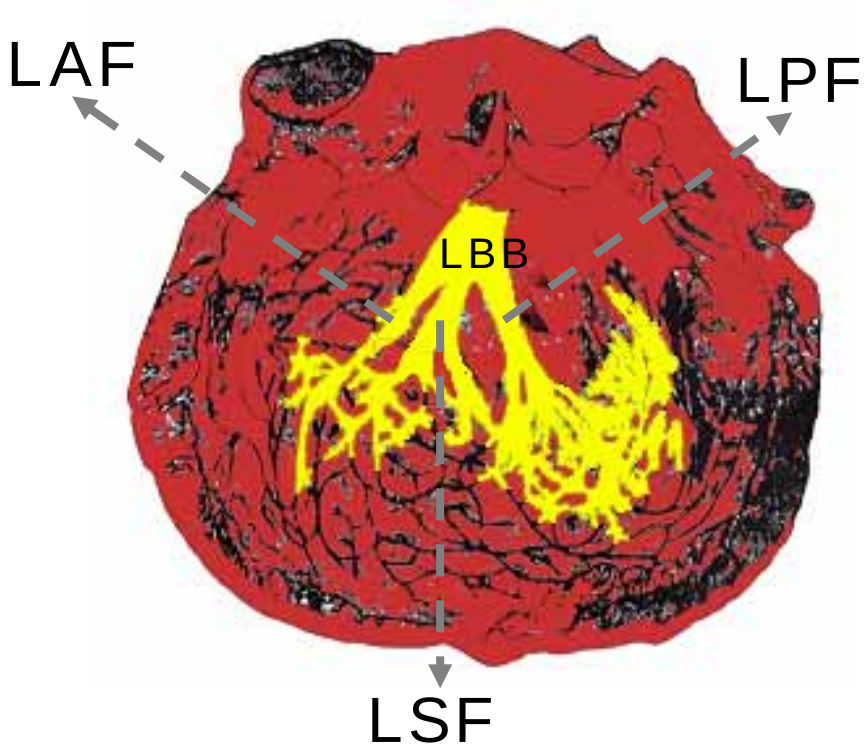
Mauricio Rosenbaum and his school "established" in the world of electrocardiography, the trifascicular concept of the intraventricular His system (RB + LAF + LPF) (1-2-3-4).

Uhley proposed (5) the tetrafascicular concept: RB + LAF + LPF + LSF.

We believe in the existence of six fascicles with electrovectocardiographic expression: LAF + LPF + LSF for the LF and three fascicles for the RB: superior or subpulmonary fascicle of the RB, middle fascicle of the RB, and inferior or postero-inferior fascicle of the RB.

1) Rosenbaum MB. J Electrocardiol 1969;2:197-206.
2) Rosembaum MB, et al. Am Heart J 1969; 78: 306-312
3) Rosembaum MB, et al. Mod Concepts Cardiovasc Dis 1970 39:141-7
4) Rosembaum MB, et al. The Hemiblocks. Tampa tracing, Florida 1970; p1
5) Uhley HN.: The Quadrifascicular Nature of the Peripheral Conduction System, in Dreifus LS, and Likoff W,.(eds.): Cardiac Arrhythmias (New York): Grune & Stratton. Inc. 1973

THE TRIFASCICULAR NATURE OF THE LEFT HIS SYSTEM



At the end of the 19th century, His described the left His system as being trifascicular (1).

Tawara (2), at the beginning of the 20th century (1906), clearly showed that the truncus of the left branch of the His bundle (LB) splits into three fascicles and not into two: left anterior fascicle (LAF), left posterior fascicle (LPF) and left septal fascicle (LSF).

1) **His, W.. JR. – Die Tatigkeit des embryonalen herzens deren Redeutung für die Lehere von der Herzbewegung beim Erwachsensn. Med. Klin. 1: 14, 1893.**
2) **Tawara S: Das Reizleitungssystsem des Saeugetierherzens: eine anatomhistologische Studie ueber die Atrioventriculaer Buendel und die Purkinjeschen Faden. Jena, Gustav Fischer; 1906.**

The left His system as Tawara considered it.

CHARACTERISTICS OF THE LEFT SEPTAL FASCICLE (LSF) OF THE HIS BUNDLE

WE WILL SUCCESSIVELY ANALYZE IN THE LSF:

1) Possible Anatomical Variations;

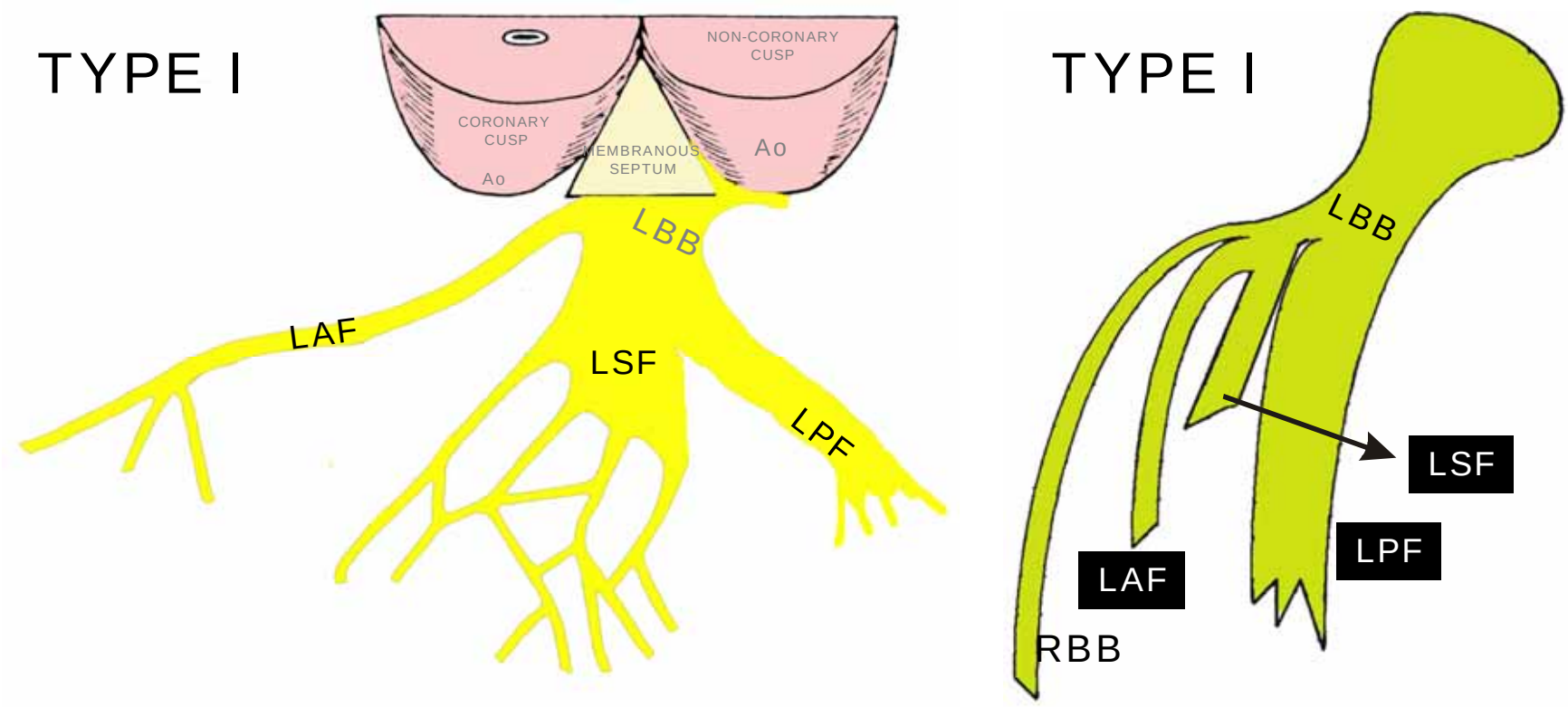
2) Conduction Velocity;

3) Location And Trajectory;

4) Blood Supply.

Characteristics to be analyzed in the left septal fascicle (LSF).

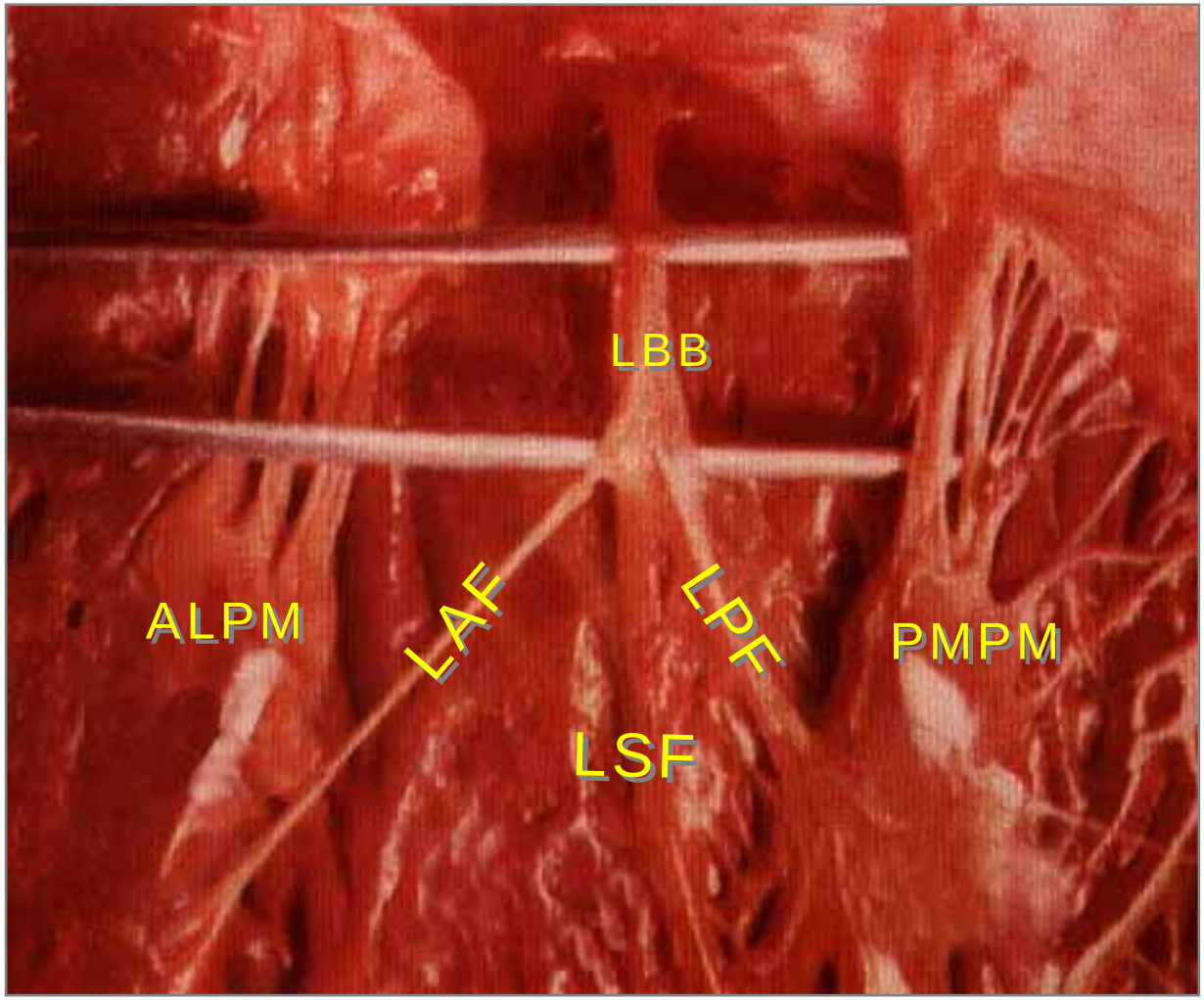
POSSIBLE ANATOMICAL VARIATIONS OF THE LSF

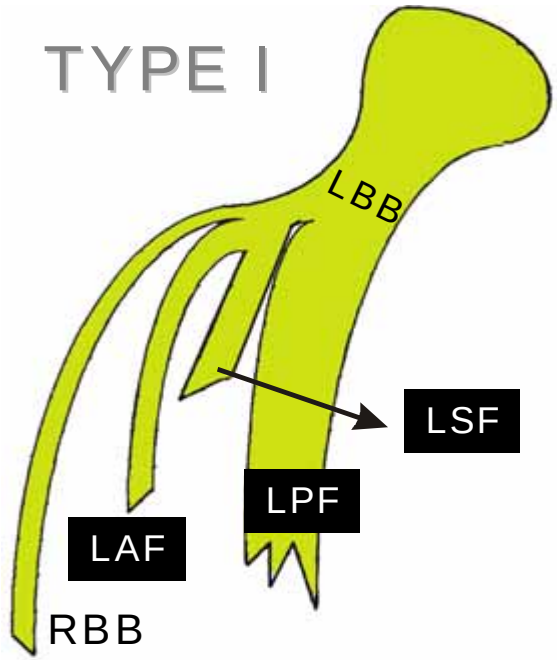


TYPE I: it is born independently from the truncus of the left branch (LB). It constitutes 65% of the cases.

LSF Type I.

INTERPRETATION OF THE TRIFASCICULAR NATURE OF THE HUMAN LEFT HIS SYSTEM





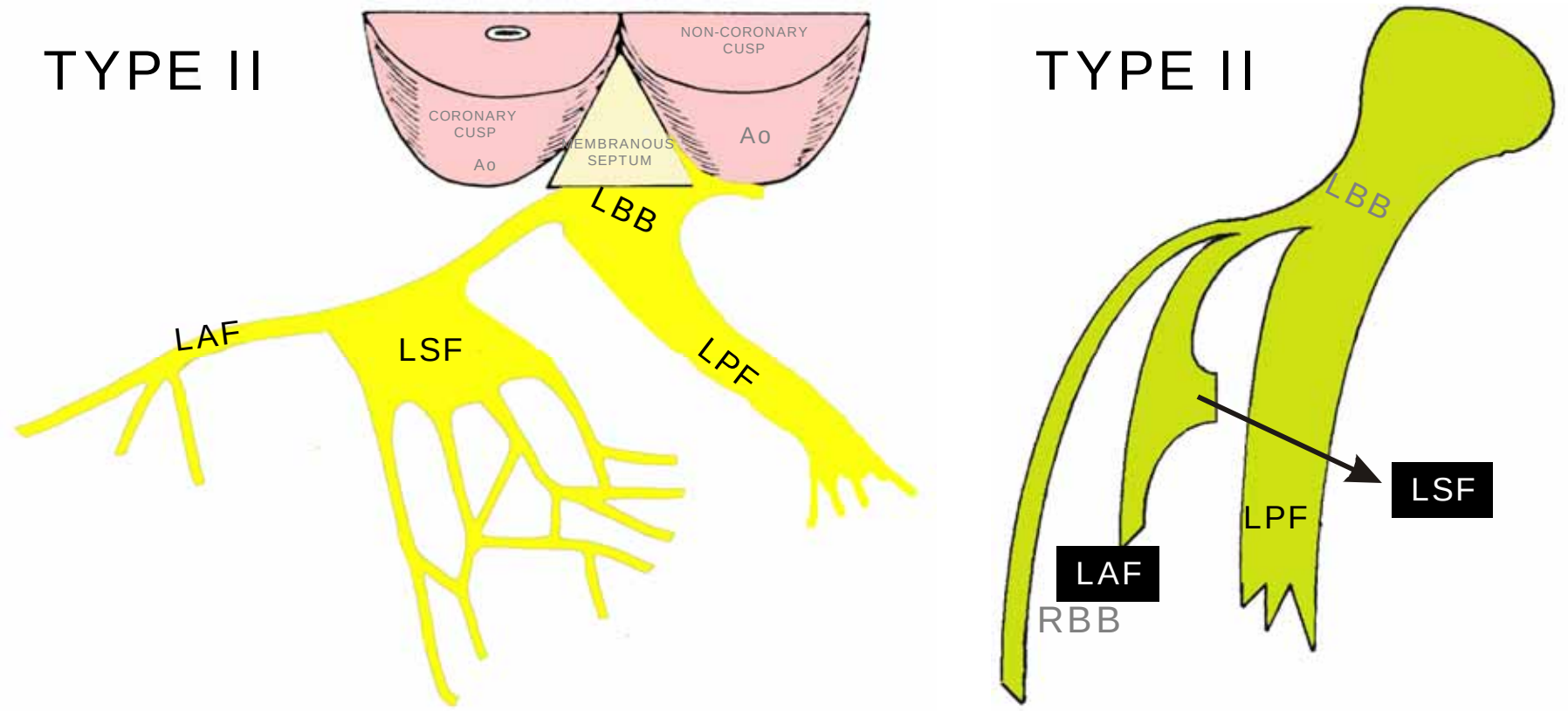
PMPM POSTERO-MEDIAL PAPILLARY MUSCLE

ALPM ANTERO-LATERAL PAPILLARY MUSCLE

In this sample of the human heart, we observe a lateral view of the left side of the interventricular septum. Undoubtedly, the LSF originates directly from the LB truncus. This variant is the most frequent one: 65% of the cases.

Sample that shows LSF Type I.

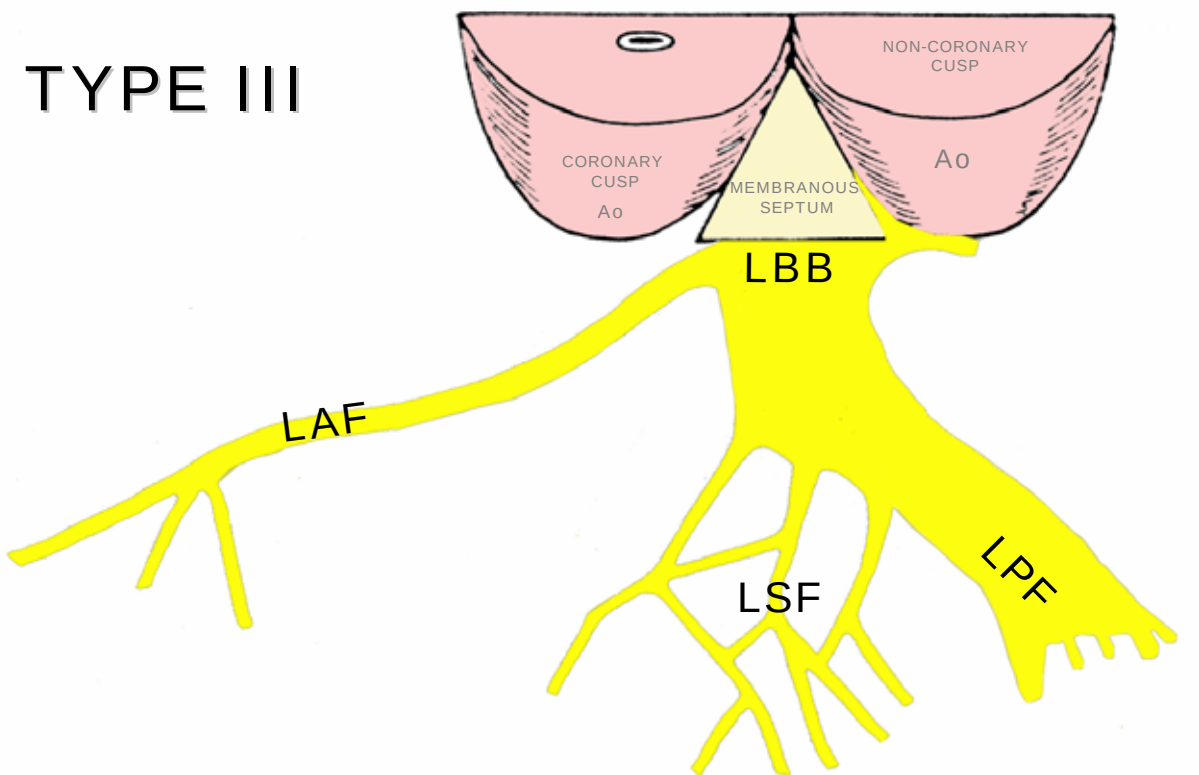
POSSIBLE ANATOMICAL VARIATIONS OF THE LSF

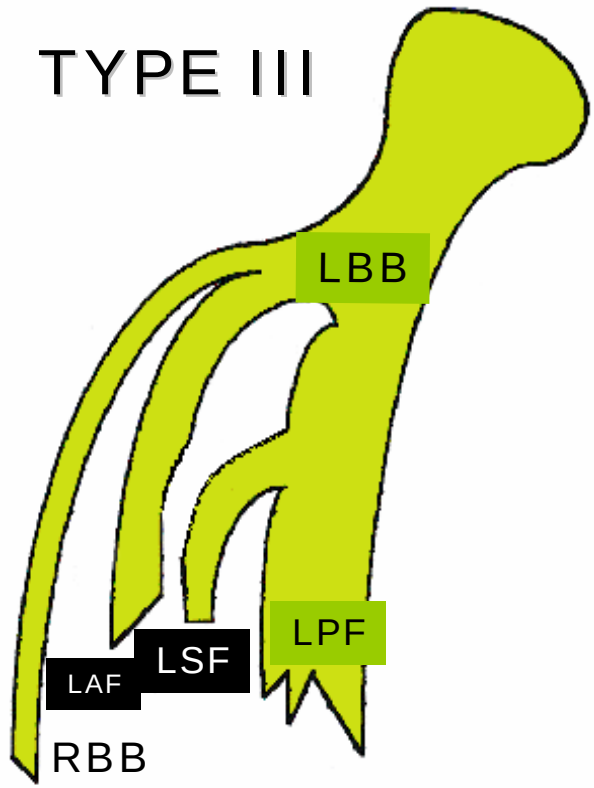


TYPE II: The LSF originates in the left anterior fascicle (LAF) of the LB.

LSF Type II.

POSSIBLE ANATOMICAL VARIATIONS OF THE LSF

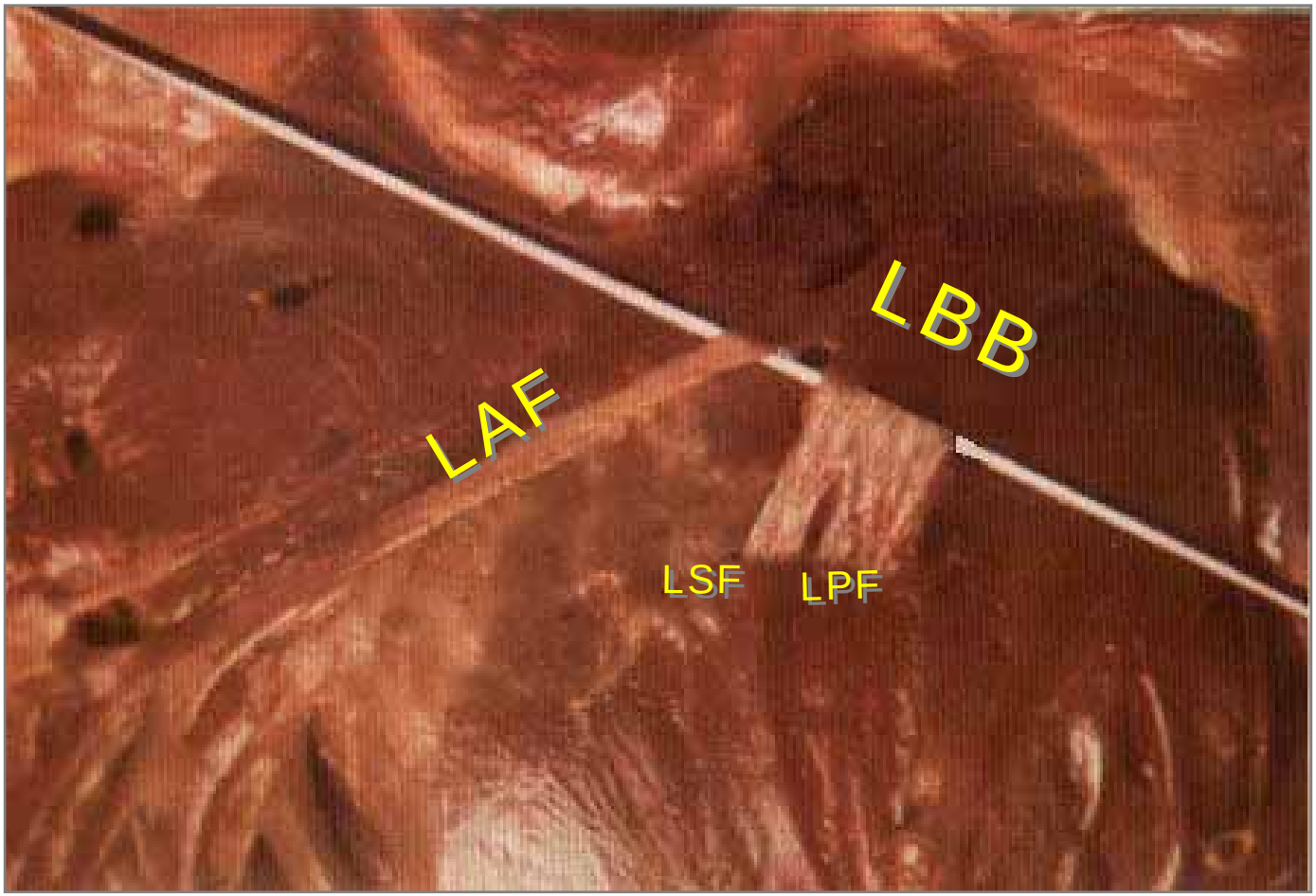


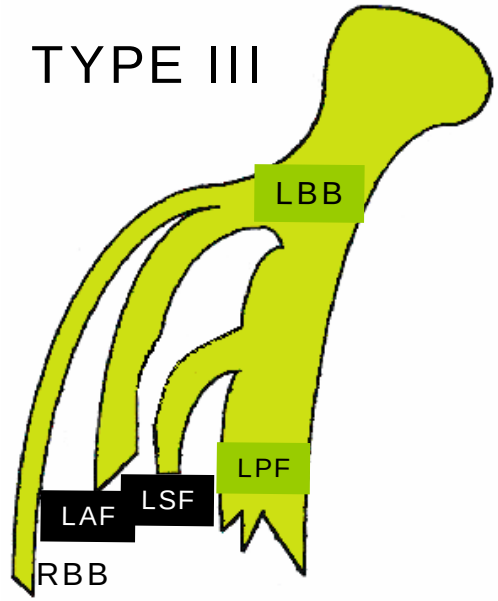


TYPE III: The LSF originates from the left posterior fascicle (LPF). Rosenbaum interpreted it as “false tendons of the LPF”.

LSF Type III.

INTERPRETATION OF THE TRIFASCICULAR NATURE OF THE HUMAN LEFT HIS SYSTEM





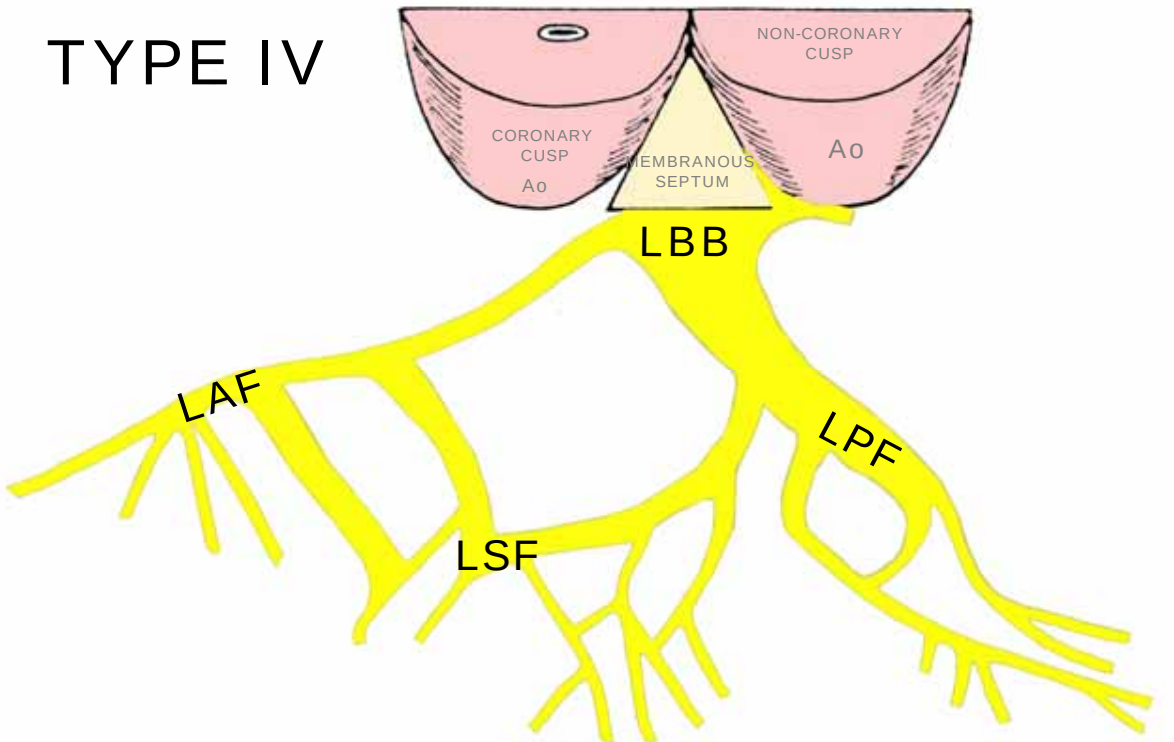
In the figure extracted from the original book by rosenbaum (1), we believe the LSF originates in the LPF (TYPE III). Rosenbaum called this “false tendons of the LPF”.

1) Rosenbaum MB, Elizari MV, Lazzari JO. Los Hemibloqueos. Editorial Paidos. Buenos Aires, 1967

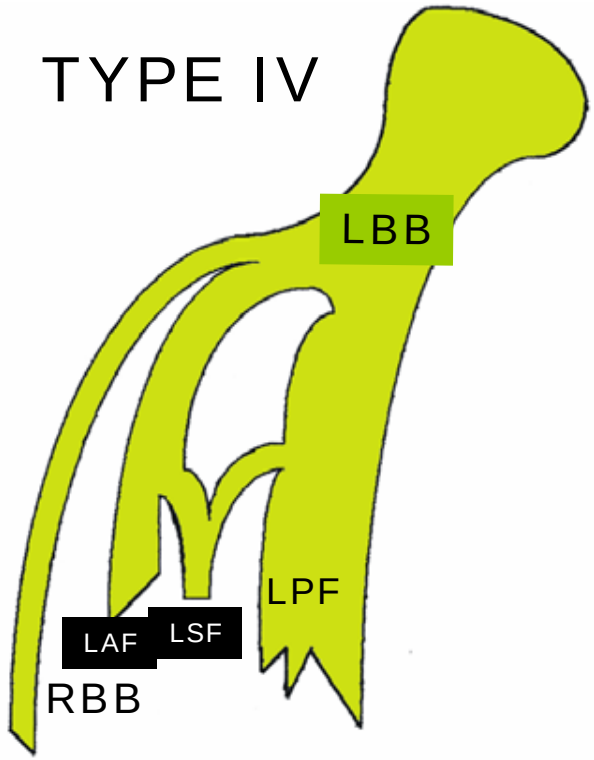
Sample that shows LSF Type III.

POSSIBLE ANATOMICAL VARIATIONS OF THE LSF

TYPE IV



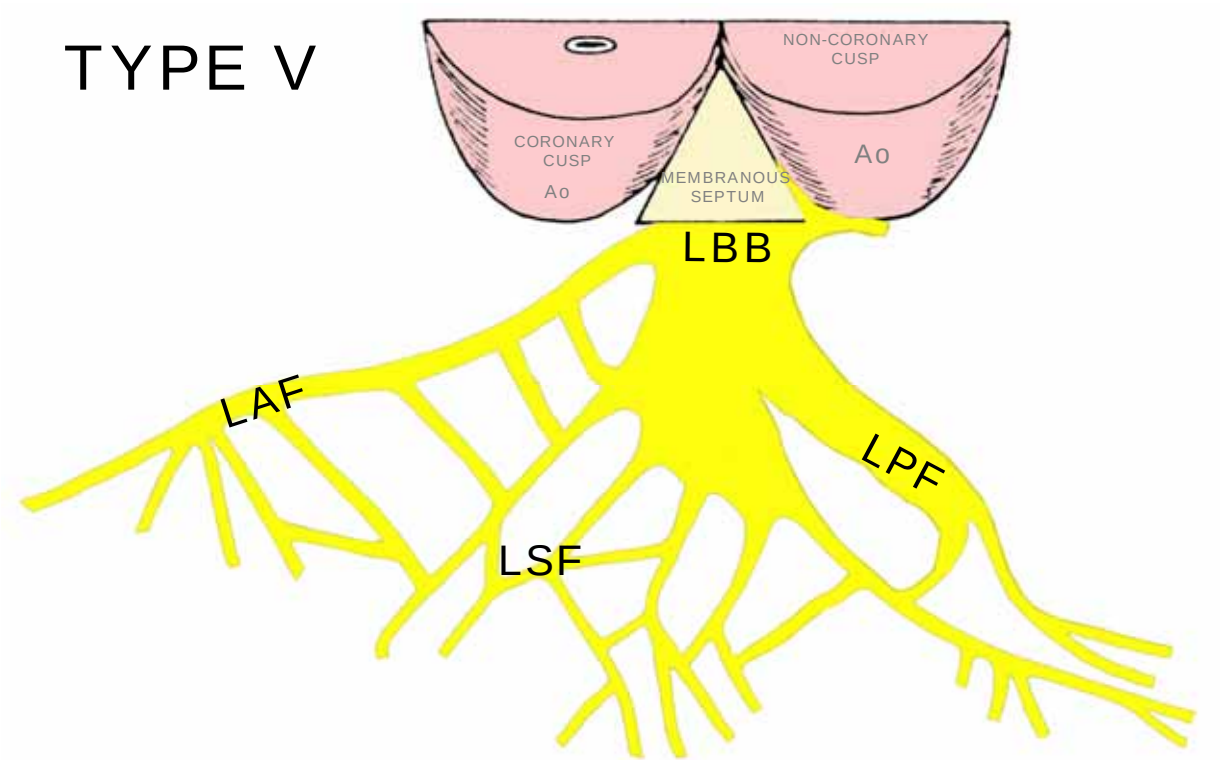
TYPE IV

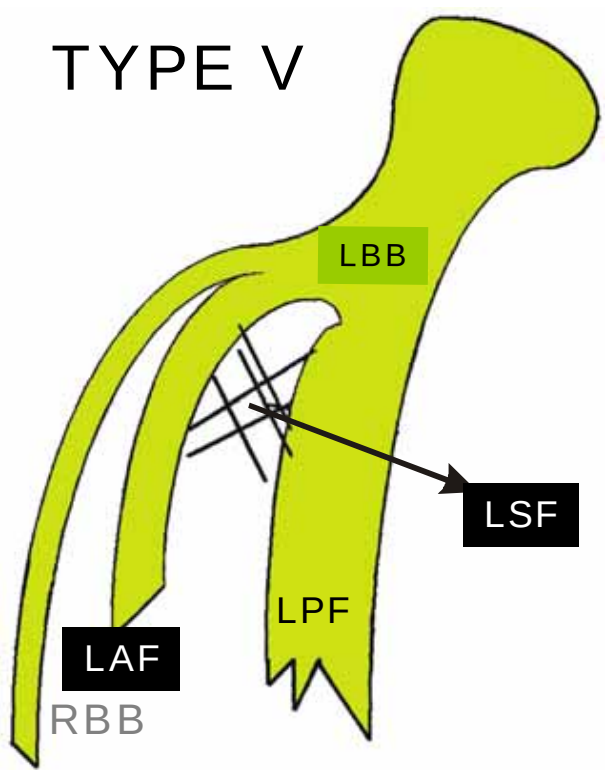


TYPE IV: the LSF is the branch of the other two fascicles of the LB: LAF and LPF.

LSF Type IV.

POSSIBLE ANATOMICAL VARIATIONS OF THE LSF

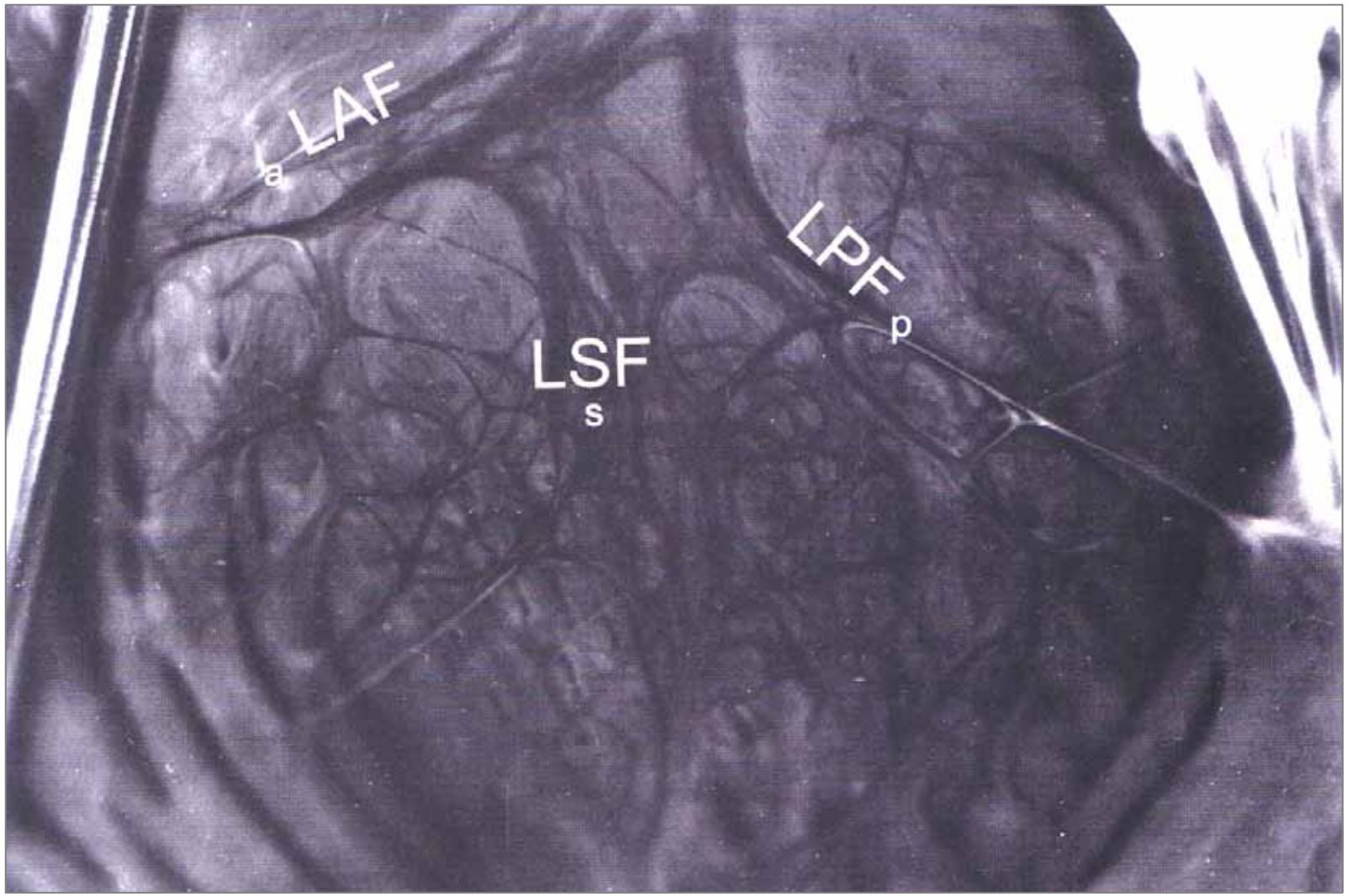




TYPE V: the LSF is a net in a fan shape, which joins both fascicles: LAF and LPF.

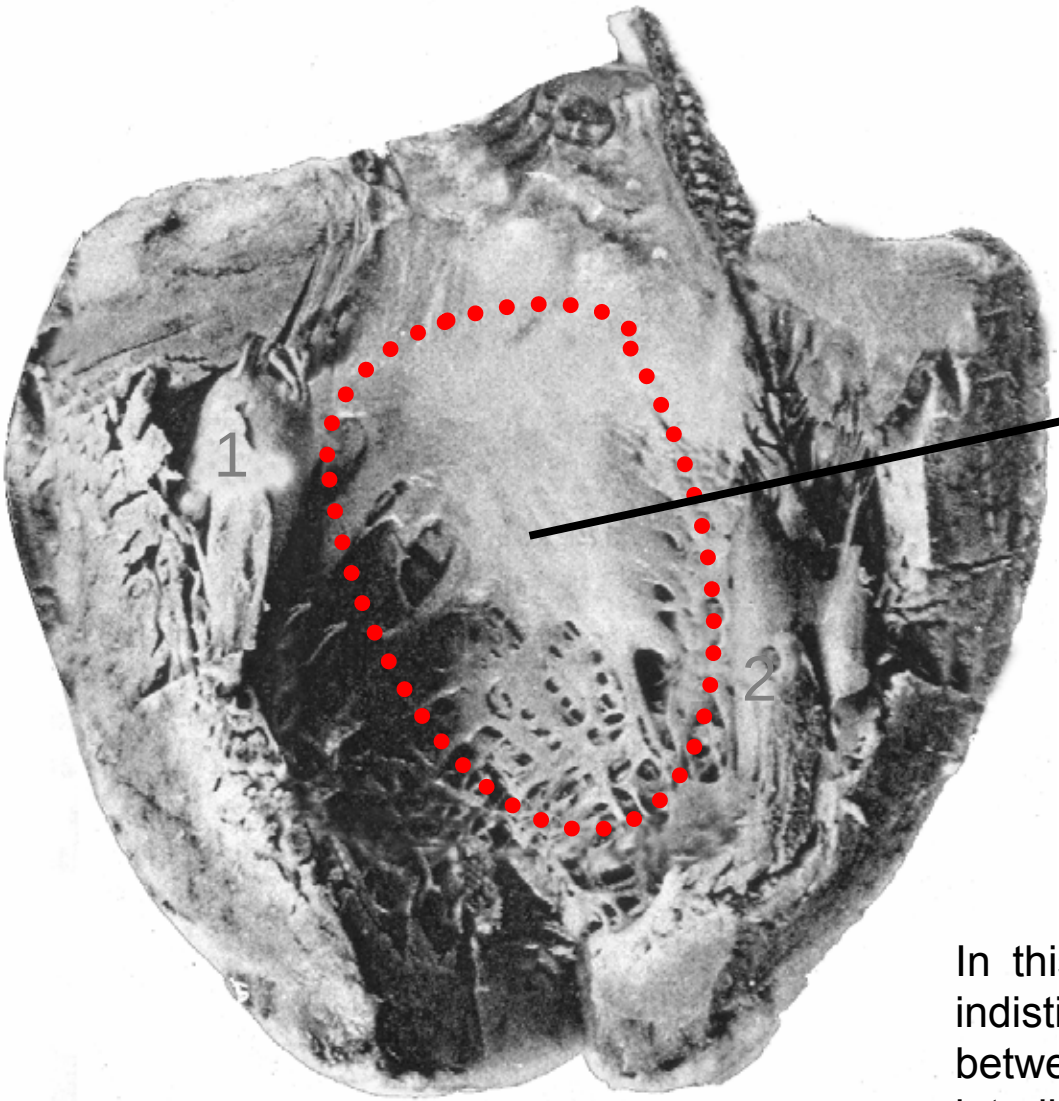
LSF Type V.

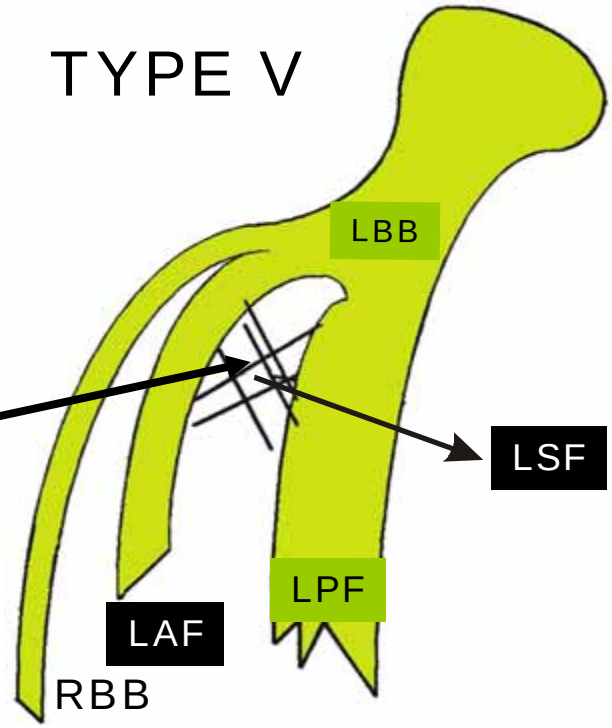
TYPE V



Photograph of the left bundle branch stained by iodine in a canine heart. In addition to the anterior (a) and posterior (p) Fibers, it shows a third, central ramification (s).

INTERPRETATION OF THE TRIFASCICULAR NATURE OF THE HUMAN LEFT HIS SYSTEM



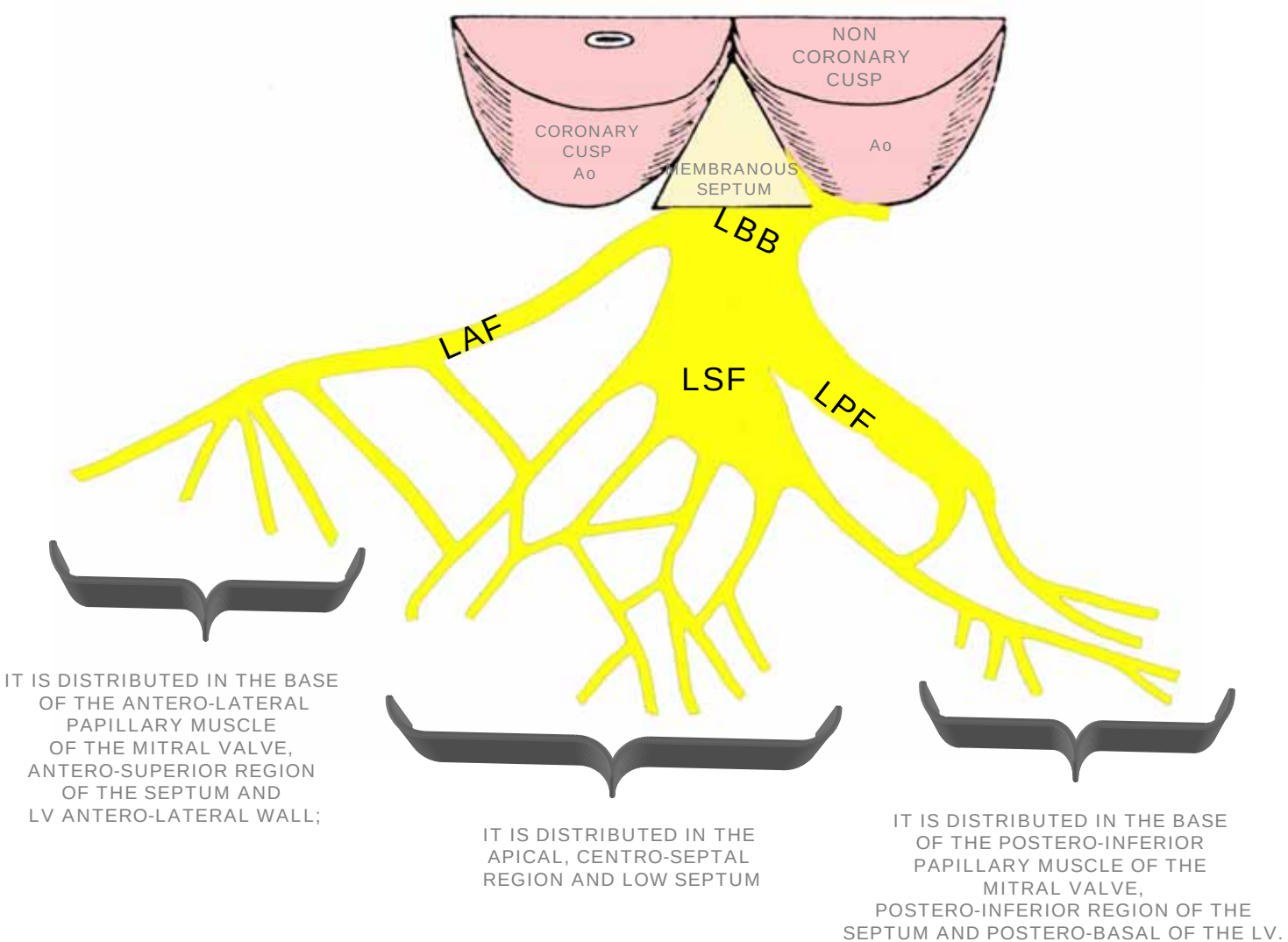


1) ANTERO-LATERAL PAPILLARY MUSCLE
2) POSTERO-MEDIAL PAPILLARY MUSCLE

In this case, the fascicles of the LB are indistinguishable, behaving as a fan between the two papillary muscles, as an interlinked net of fibers with the shape of a fan.

Sample that shows LSF Type IV.

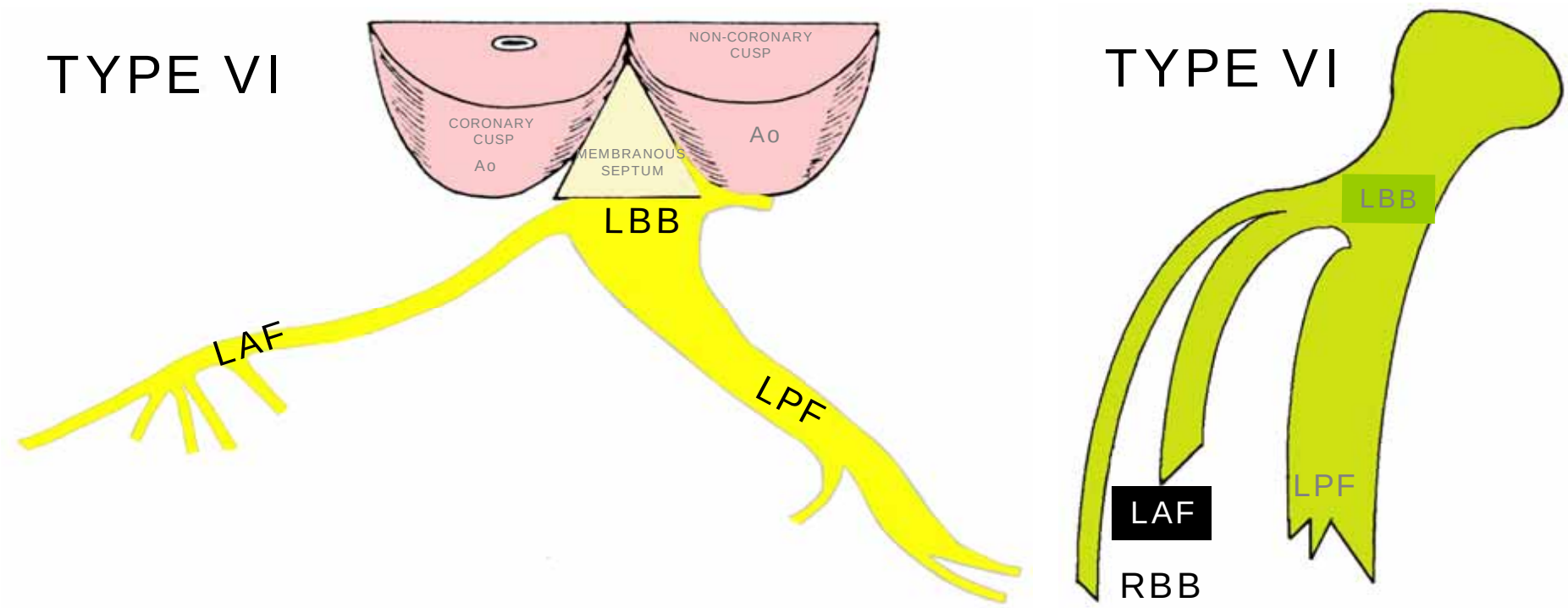
THE TRIFASCICULAR CONCEPT OF THE LEFT HIS SYSTEM AND THE VENTRICULAR DISTRIBUTION OF ITS FASCICLES



Modified from Hecht HH, et al. Am J Cardiol 1973; 31:232-244

Distribution of the three fascicles of the left branch.

POSSIBLE ANATOMICAL VARIATIONS OF THE LSF



TYPE VI: There is no LSF. 15% of the cases: bifascicular left his system. This percentage rises to 40% in the histopathological work by Kulbertus. (1)

1) Kulbertus HE. Adv Cardiol. 1975;14:126-135.

LSF Type VI. When the LSF does not exist.

POSSIBLE ANATOMICAL VARIATIONS OF THE LSF

CONCLUSIONS ON THE CONTROVERSY OF THE BIFASCICULAR OR TRIFASCICULAR NATURE OF THE HUMAN LEFT HIS FASCICLE

Taking as basis the commented aspects, we conclude that in most cases, the left his system is predominantly trifascicular and not bifascicular. consequently, the term “hemiblock”, established by Rosenbaum and his school (1-2) is inappropriate.

We believe that the following thought by Fernando de Pádua, researcher of the portuguese school, is extremely appropriate (3-4-5):

“IF HEMIBLOCKS DO EXIST, THEY ARE ONLY TWO - IF A THIRD ONE IS POSTULATED, HEMIBLOCKS DO NOT EXIST !”

1) Rosenbaum, M B, Elizari M V and Lazzari, J O: Los Hemibloqueos. Paidos, Buenos Aires, 1968

2) Rosembaum MB, Elizari MV, Lazzari JO: The Hemiblocks: Diagnostic criteria and clinical significance. Mod Concepts Cardiovsc Dis 1970 39:141

3) De Pádua F, Lopes VM, Reis DD, et al. - O hemibloqueio esquerdo mediano - Uma entidade discutível. Bol Soc Port Cardiol 1976

4) De Pádua F, Reis DD, Lopes VM, et al. - Left median hemiblock - a chimera? In: Rijlant P; Kornreich F, eds. 3rs Int. Congr. Electrocardiology. (17th Int. Symp. Vectorcardiography). Brussels, 1976

5) De Pádua F. Bloqueios fasciculares – os hemibloqueos em questão – Rev Port Clin Terap 1977; 3:199-200

Reflections on the tri or bifascicular nature of the left His system.