

Atypical ECG in young healthy woman (19 years-old)

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Case report: Young Caucasian woman without complaint. Irrelevant personal and familiar history.

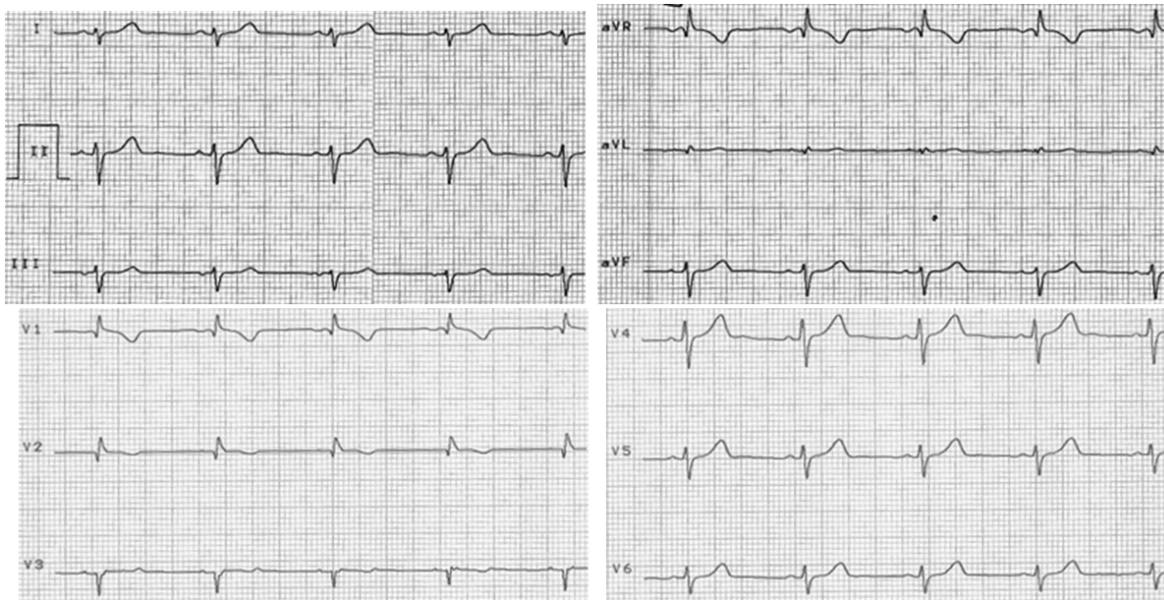
Physical examination: nothing of note.

ECG performed for clinical evaluation before cosmetic surgery.

Which is the ECG diagnosis? And why?

Observation: We are sure that the ECG was performed correctly (precordial electrodes in appropriate positions).

Figure 1



Colleague's opinions

This ECG is suggestive of primum ASD defect.

A detailed analysis discussing "differential diagnosis" and how you might differentiate the possibilities would be useful.

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Dear colleagues.

SISIISIII and SII>SIII. Frontal plane QRS axis deviation to the right superior quadrant. qR in aVR. qR with T inversions in V1-V3. Epsilon wave in V3?

Right end-conduction delay type 1A, maybe associated with ARVC??

Best regards
Kjell Nikus
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A primera impresión pienso lo siguiente.

El trazado puede corresponder a un paciente sano con corazón dextrorotado, y retardo de la activación ventricular derecha, no necesariamente un bloqueo de rama derecha troncular.

Dudo mucho que las derivaciones V1-V2-V3 estén colocadas en su lugar correcto. La onda P es negativa en V1 y plana en V2, cuando normal en el plano frontal.

Rogaría ver un ECG con V1-V6 en lugar correcto, y V1-V3, 2-3 centímetros más alto. Si no hay cambios podría pensarse en algo patológico pero no creo que sea DAVD (no hay onda Epsilon y no hay T negativa evidente simétrica en V1-V4).

Adjunto un artículo original mío escrito en el 1987 sobre la morfología S1-S2-S3.

Con todo afecto.

A. Bayés de Luna

Retardo final de conduccion en la rama derecha?

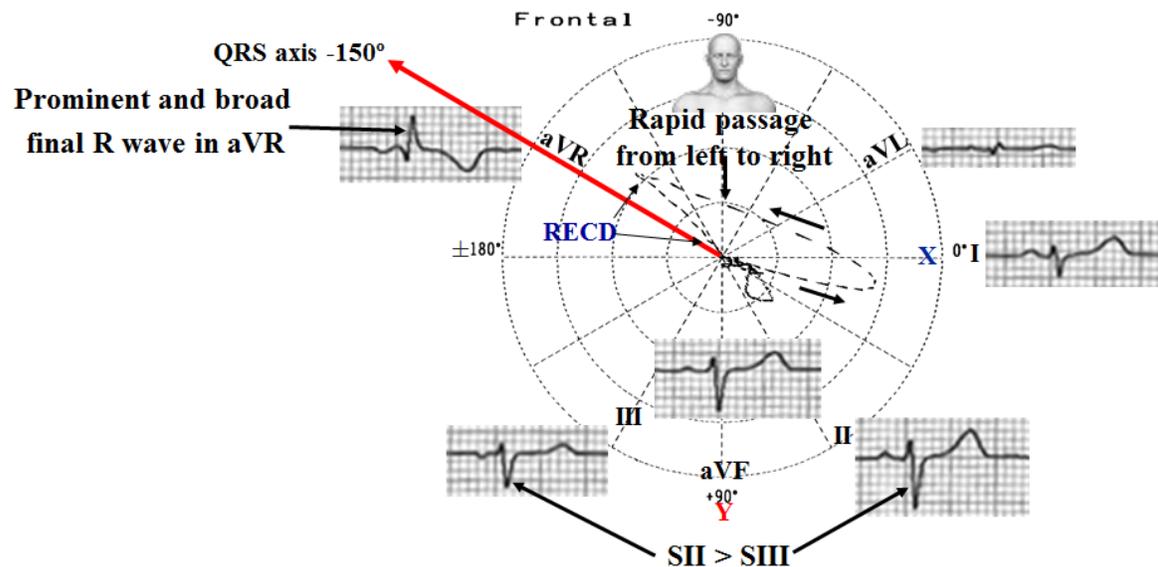
Marcelo Carreon

Final diagnosis

Clinical diagnosis: Normal heart

ECG diagnosis Superior Fascicular Right Bundle Branch Block (SFRBBB)

Figure 2 - ECG/VCG correlation on Frontal Plane



Extreme right axis deviation: QRS axis between -90° and $\pm 180^\circ$ (AKA “Northwest Axis”): no man's land. This QRS axis deviation is observed in: emphysema, hyperkalaemia, lead transposition, artificial cardiac pacing, ventricular tachycardia and right bundle branch fascicular block on free wall of the right ventricle. It is indicative of slow conduction (slow dromotropism) on territory of right ventricular outflow tract (RVOT). In this region runs the superior or subpulmonary fascicle of the right bundle branch. In this case, the QRS loop has a counterclockwise rotation, very similar to observed in left anterior fascicular block (LAFB), but has several important differences between both dromotropic disorders. See next slide.

Table 1 - Differential diagnosis between Superior Fascicular Right Bundle Branch Block (SFRBBB) and Left Anterior Fascicular Block (LAFB) of Left Bundle Branch

	SFRBB	LAFB
Depth of S wave in II and III	SII > SIII (inconstant)	SIII > SII (inconstant)
I and aVL pattern	Rs or rs	qR
Prominent and broad final R wave in aVR	Present and characteristic: QR or qR.	Absent: Qr or QS
Vector of initial 10 to 20 ms	Downward and to the left (inconstant)	Downward and to the right. (inconstant)
Rapid passage from left to right	Yes	No
Triphasic or biphasic pattern in V ₁ or V ₁ and V ₂	Very frequent	Possible. Final R' wave or r' wave of V ₂ is greater than in V ₃ R and V ₄ R, indicating that the final forces are heading predominantly to the left.

Figure 3

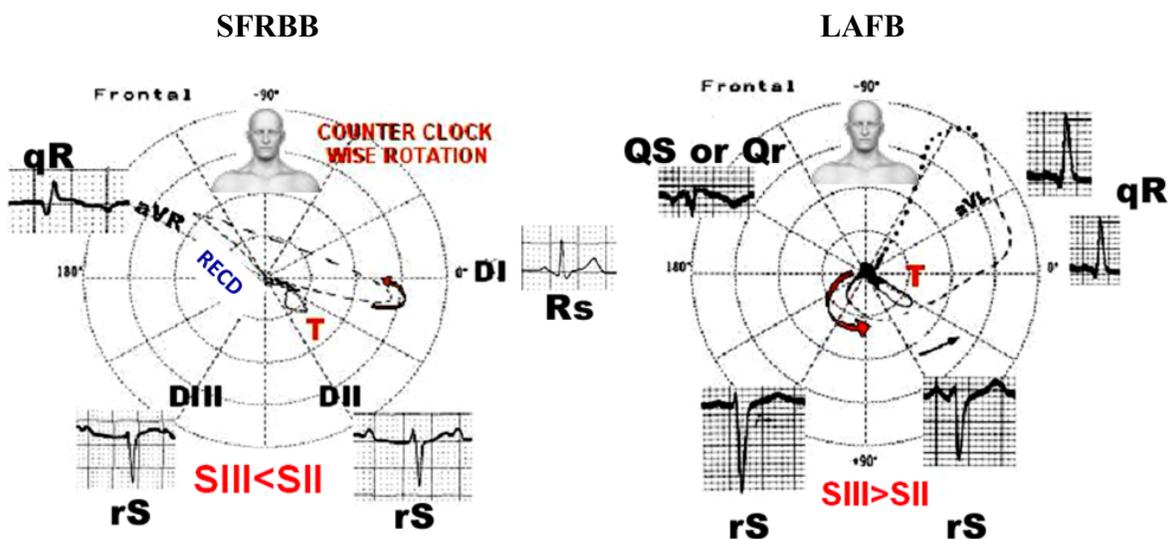
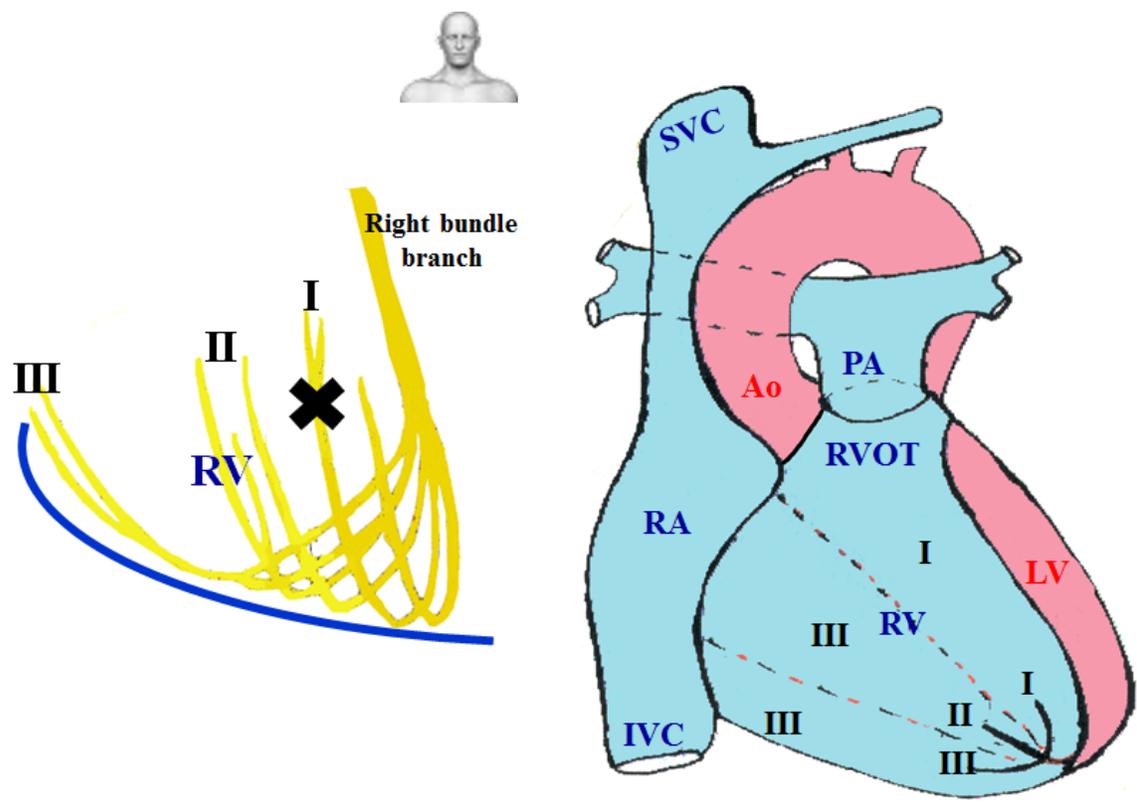


Figure 4 - Distribution of the three fascicles of the Right Bundle Branch in the RV free wall



I - Territory of superior or subpulmonary fascicle of the RBB

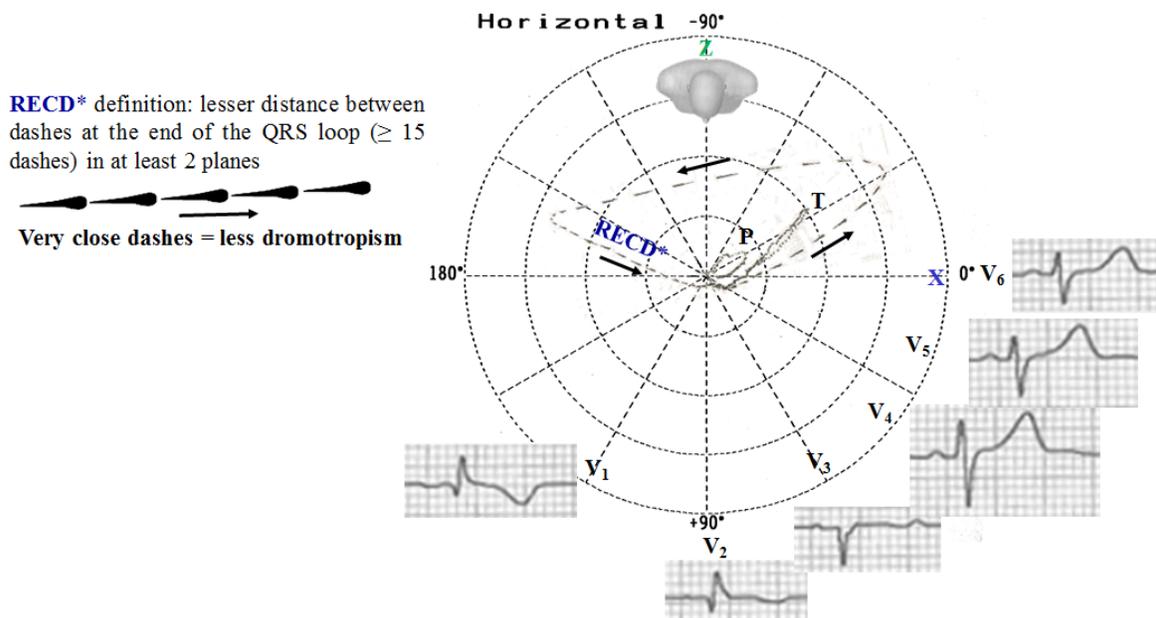
II – Territory of inferior or posteroinferior fascicle of the RBB

III – Territory of middle fascicle of the RBB

RVOT – Right Ventricular Outflow Tract

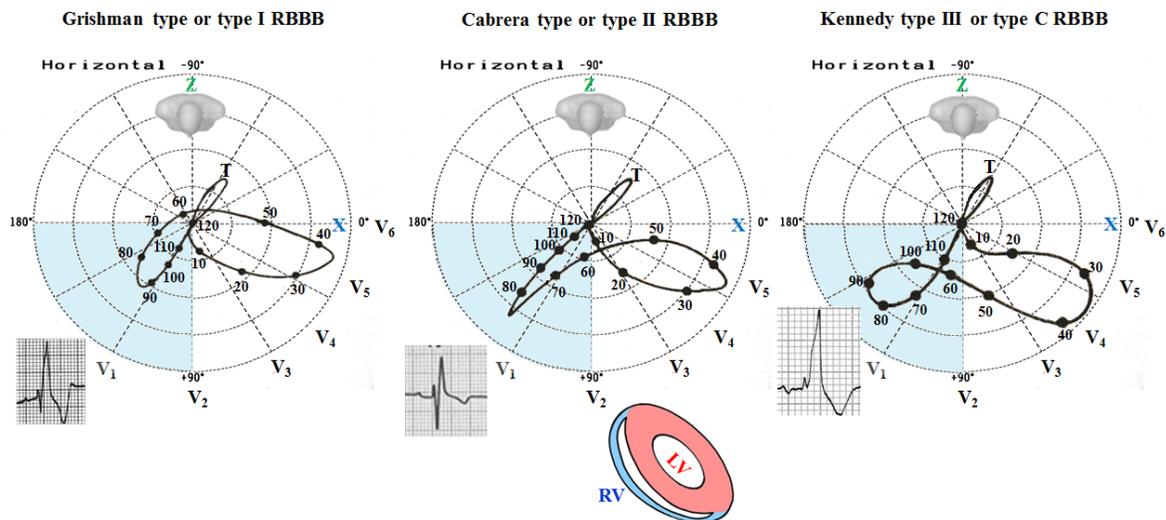
✘ Blocked zone

Figure 5 - ECG/VCG correlation in the Horizontal Plane



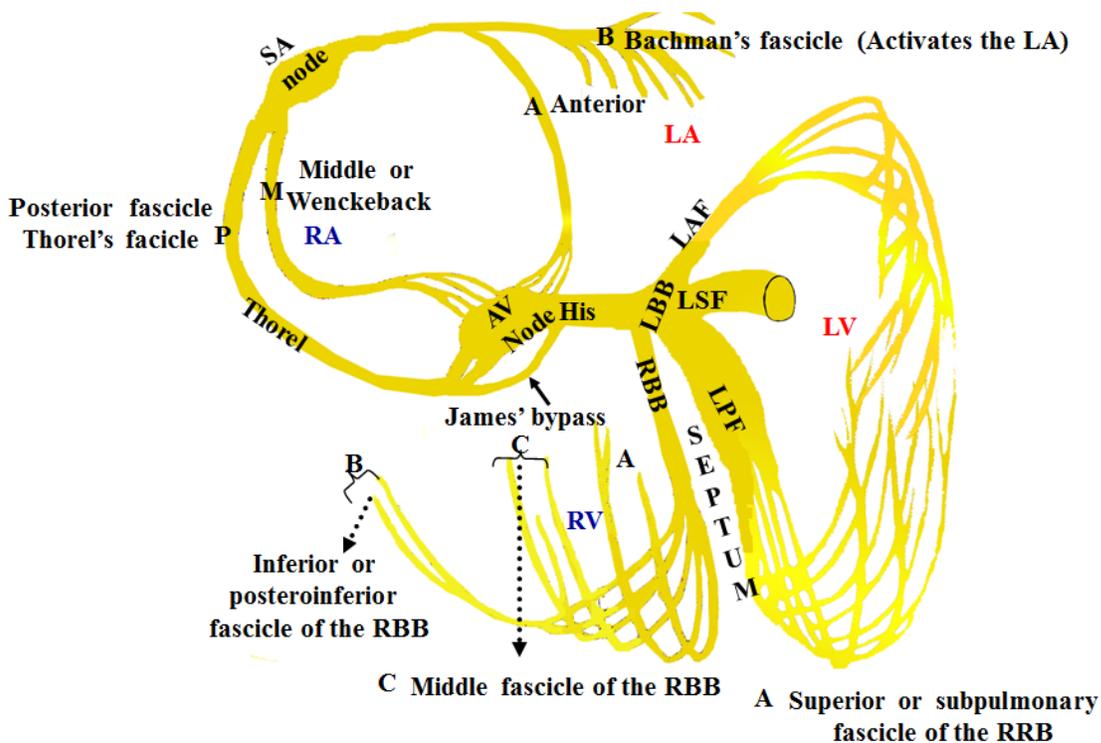
qR in V1-V2, rS from V3 to V4, pseudo anteroseptal infarction associated with incomplete RBBB. 20ms vector directed to left, the QRS loop is dislocated to back and rightward and right end conduction delay(**RECD***) on posterior right quadrant. **RECD*** means: lesser distance between the dashes at the end of QRS loop (≥ 15 dashes) in at least two planes. In troncular or predivisional RBB, the RECD is located on anterior right quadrant. See next figure.

Figure 6 - Troncular RBBB QRS and T loops in the Horizontal Plane. Note **RECD (light blue) located on anterior right quadrant after 60 ms**



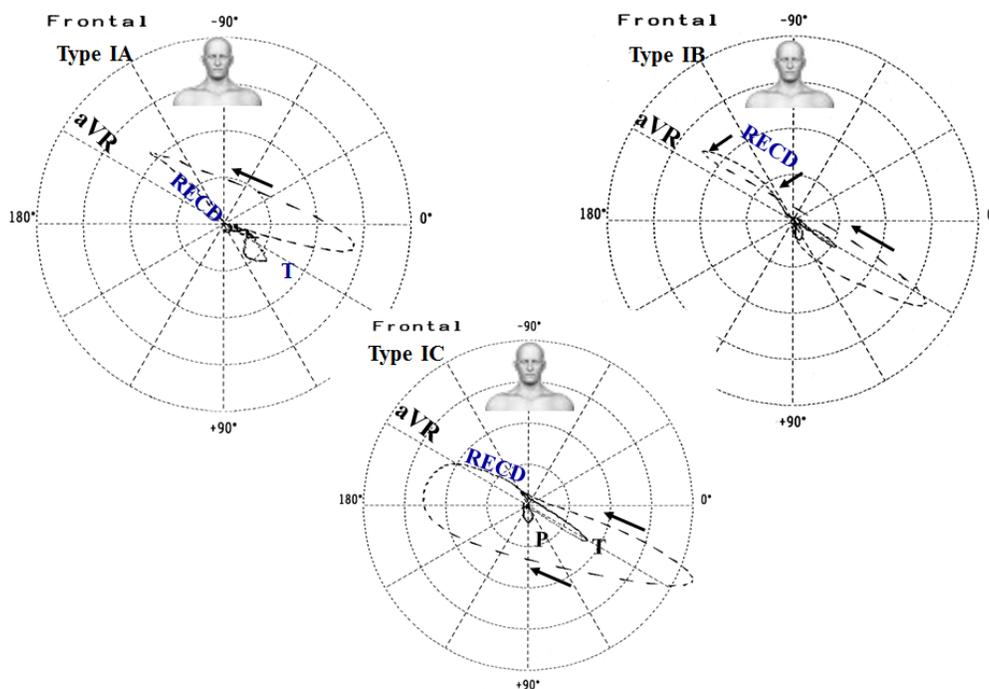
In the 3 troncular RBBB types, RECD is always located on right anterior quadrant with characteristic finger-like terminal apêndix.

Figure 7 - The hexafascicular concept of intraventricular conduction system



In this figure we see the SA node, atrial internodal bundles (anterior, middle and posterior), AV node, His bundle and its divisions (3 left and 3 right).

Figure 8 - VCG proposal classification of the superior fascicular right bundle branch block (SFRBBB) according to the VCG QRS loop pattern in the frontal plane



RECD located on top right quadrant near aVR lead (-150°). Only the type IA has differential diagnosis with LAFB because both dromotropic disorders have Counter clockwise rotation.

Clinical significance

What are the so called right end conduction delay (**RECD**) or fascicular, parietal or partial Purkinje right bundle branch blocks? Answer: these are the electrovectorcardiographic changes, secondary to physiological delay or to true dromotropic disorders in the territory of one of the three fascicles of the right branch, in isolation in the RV free wall. See next slide. Its clinical significance and interest lies in the fact that:

- They may be confused with left fascicular blocks: Left Anterior Fascicular Block (LAFB) and Left Posterior Fascicular Block (LPFB);
- They may be confused with electrically inactive areas (pseudo myocardial infarction) both in the anterior and the inferior walls.
- They may represent the electro-veclocardiographic pattern of Brugada syndrome (1) and one subpopulation of Arrhythmogenic Right Ventricular Dysplasia (ARVD/C).

Why not ostium primum atrial septal defect (OP-ASD)?

Atrial Septal Defects (ASD)

Figure 9 shows localizations of the main ASD and CS-ASD.

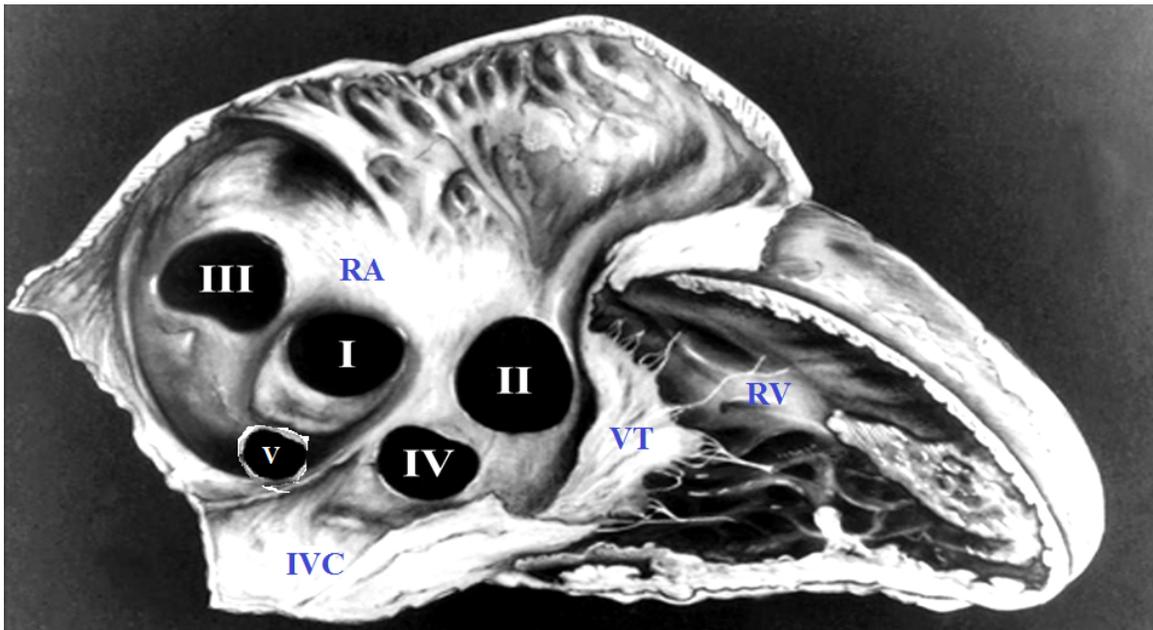


Figure 9: I) Ostium Secundum (OS-ASD); II) Ostium Primum (OP-ASD); III) Sinus Venosus (SV-ASD); IV) Coronary Sinus (CS-ASD); Inferior Vena Cave (IVC); Right Atrium (RA); Right Ventricle (RV).

Figure 10 - Possible anatomical component of OP-ASD

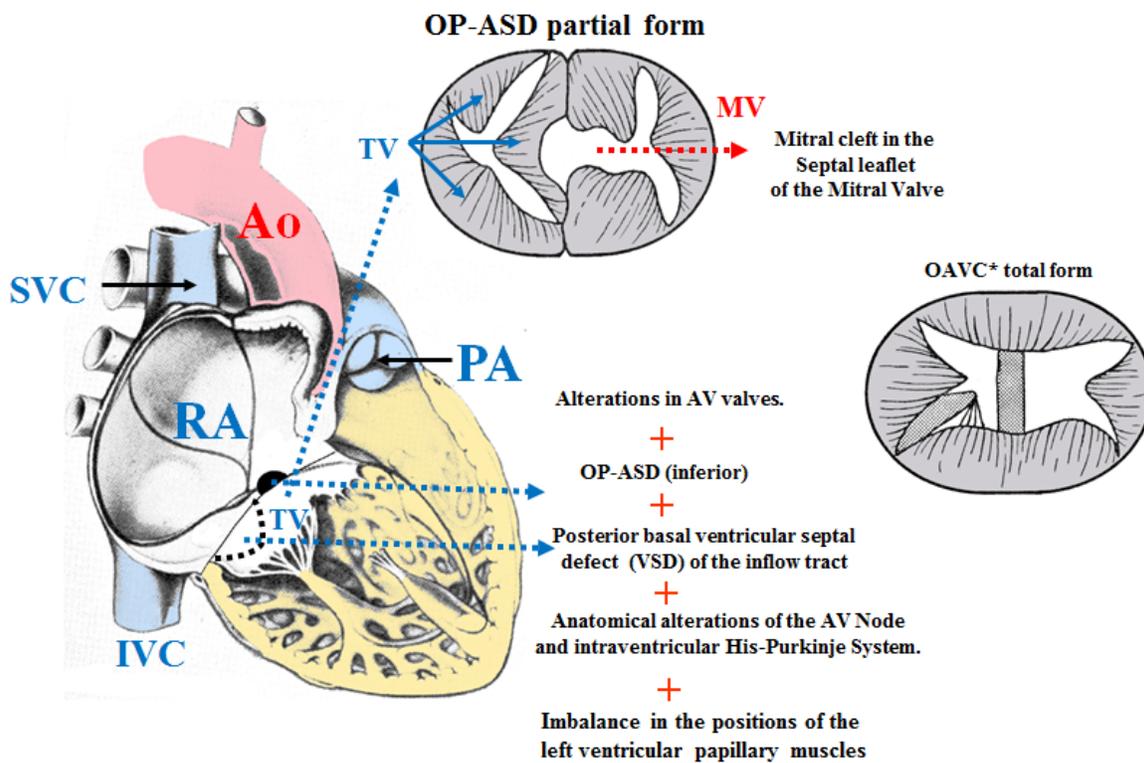


Figure 10 shows the main features in endocardial cushion defects or OP-ASD, of the endocardial cushion defect type:

- 1) Alterations in AV valve (cleft);
- 2) Low atrial septal defect ASD (OP-ASD);
- 3) Posterior basal ventricular septal defect (VSD) of the inflow tract and,
- 4) Anatomical alterations of the AV Node and intraventricular His-Purkinje System
- 5) Imbalance in the positions of the left ventricular papillary muscles. There are two major forms: ostium primum in its partial form and ostium atrium ventricularis comunis.

Hypothesis that may explain the extreme left axis QRS deviation in OP-ASD

There are three hypothesis:

- I) Anatomical alterations of the AV Node and intraventricular His-Purkinje System
- II) Imbalance in the positions of the left ventricular papillary muscles
- III) Eclectic theory

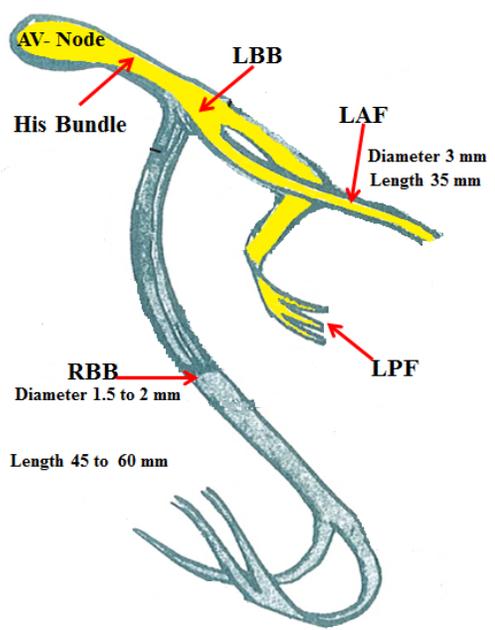
I) Anatomical alterations of the AV node and intraventricular His-Purkinje System

In endocardial cushion defects the compact AV node is inferoposteriorly displaced outside Koch's triangle adjacent to where posterior rims of atrial and ventricular septae unite (3). The His bundle extends along the lower rim of the ventricular septum. This inferior course and relative hypoplastic and longer (diameter 1mm and length 70 mm) of the left anterior fascicle gives rise to the superior QRS axis typical of endocardial cushion defect (4). In normal conditions left anterior fascicle has 3 mm diameter and 45 mm length. Additionally, an abnormally short posteroinferior fascicle causes early activation of the posterobasal region of the left ventricle with **posteromedial papillary muscle** further from the septum (5).

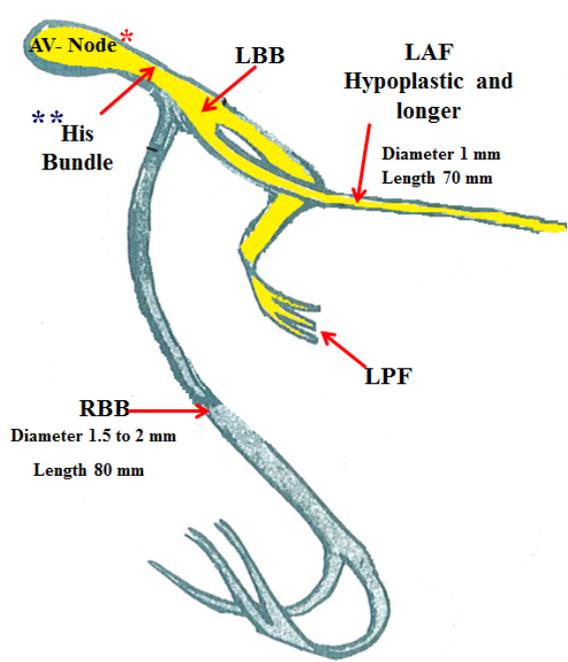
Figures 11, 12 and 13 show the anatomical alterations of the AV node and intraventricular His-Purkinje System (figure 11) the normal positions of left papillary muscles (figure 12) and the imbalance in the positions of the left ventricular papillary muscles of mitral valve (anteromedial and posterolateral).

Figure 11 - Anatomical alterations of the AV Node and intraventricular His-Purkinje System

NORMAL INTRAVENTRICULAR CONDUCTION SYSTEM



INTRAVENTRICULAR CONDUCTION SYSTEM IN ENDOCARDIAL CUSHION DEFECTS



II) Imbalance in the positions of the left ventricular papillary muscles

Another hypothesis maintains that patients with ostium primum atrial septal defect (OP-ASD) have an imbalance in the positions of the left ventricular papillary muscles compared with healthy subjects, and that this anatomic imbalance correlates with left deviation of the QRS axis. The function and contraction pattern of the heart is best predicted when cardiac anatomy is considered together with its electrical activation sequence. Understanding the electroanatomic relationships is essential for discovering the developmental relationships between the conduction system and heart structures. Left deviation of the QRS axis is typically present in patients with OP-ASD. However, the pathophysiology of this phenomenon is not understood. Thirty-five patients with OP-ASD and 35 healthy subjects were included in the study. Echocardiographic images were used to determine the papillary muscle positions. A ECG was used to determine the QRS axis in the frontal plane in both patients and healthy subjects. An imbalance between papillary muscle positions in OP-ASD patients was defined as the position of the **anterolateral papillary muscle** closer to the septum and/or the position of the **posteromedial papillary muscle** further from the septum compared with the position of the papillary muscles in healthy subjects. The figure shows normal position of LV. Figure 12 shows the normal position of left ventricle papillary muscles.

Figure 12 - Normal position of left ventricle papillary muscles

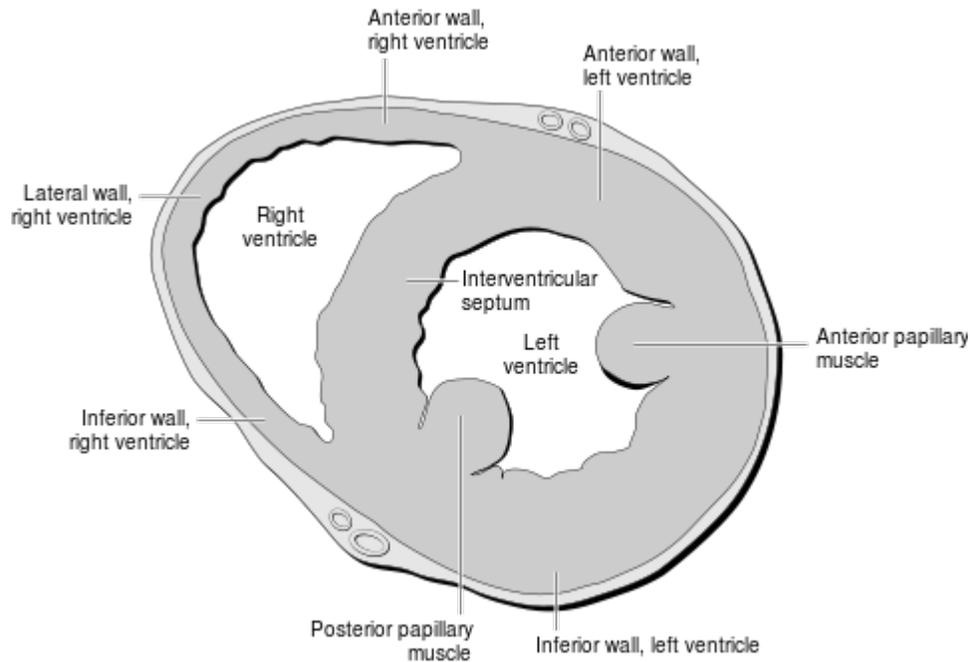
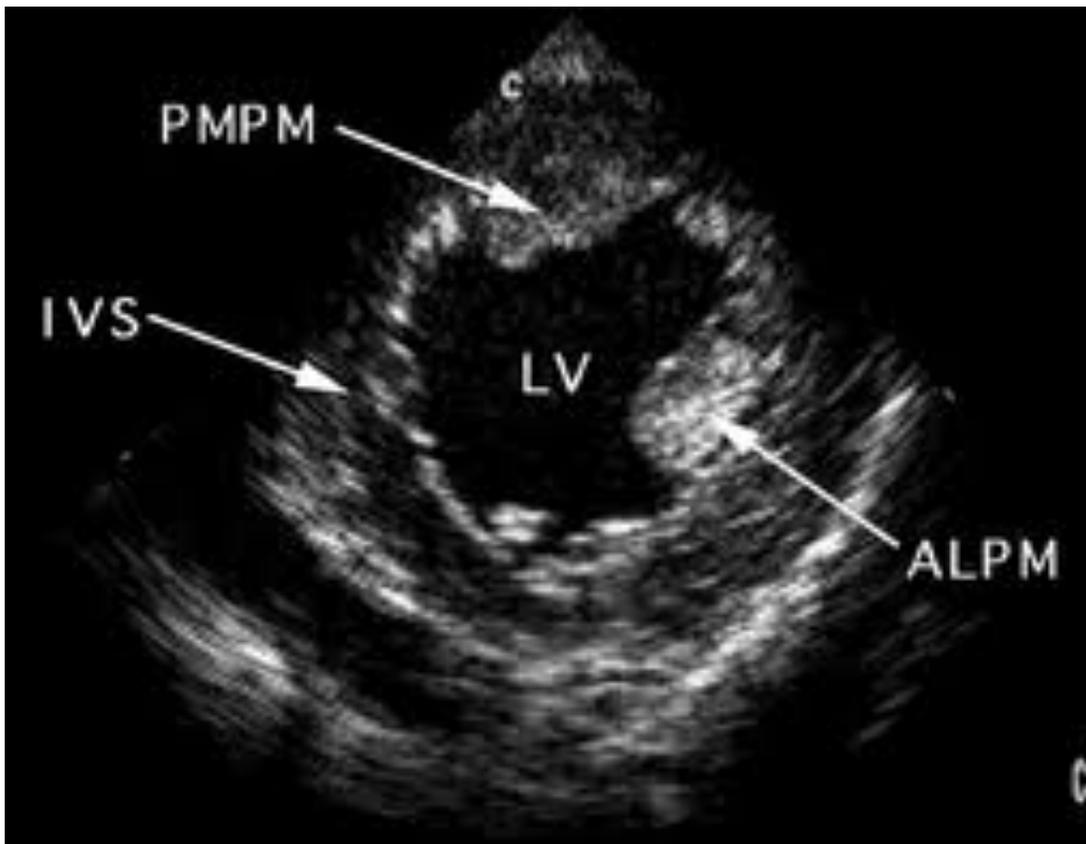


Figure 13 - Imbalance in the positions of the left ventricular papillary muscles



In OP-ASD patients compared with control subjects, there was significant imbalance in the positions of the papillary muscles. The imbalance of papillary muscles correlated with deviation of the QRS on FP. Abnormality in the position of the papillary muscles changes continuously with the abnormality of the QRS axis. Understanding the electroanatomic relationships provides important insight into developmental relationships between the conduction system and the trabecular structures in OP-ASD patients. These results may provide insights in understanding the continuity of OP-ASD abnormality, in estimating the best surgical approach, and predicting the prognosis of OP-ASD patients (6).

Eclectic hypothesis

Table 2 shows anatomical differences between healthy subject and endocardial cushion defects. This difference together explain the extreme left QRS axis deviation observed in OP-ASD

Table 2

	AV compact node and intraventricular conduction system in healthy individuals	Compact AVnode and intraventricular conduction System in Endocardial Cushion Defect
Compact AV node position	It is anterior and superior related compact AV node of Endocardial Cushion Defect	It is inferiorly and posteriorly displaced outside Koch's triangle
His bundle	Longer related His bundle of Endocardial Cushion Defect	Shorter and shifted backward
Left anterior fascicle diameter	3 mm	1 mm
Left anterior fascicle length	45 mm	65 mm
Anterolateral papillary muscle position	Further septum	Closer to the septum
Posteromedial papillary muscle position	Closer from the septum compared with the position of the papillary muscles in cushion defects .	Further from the septum compared with the position of the papillary muscles in healthy subjects.

Association of these entire factors could explain the extreme left axis deviation in OP-ASD and endocardial cushion defects.

Characteriristic of the ECG in Endocardial Cushion Defects

On ECG extreme left superior axis deviation is the rule in ostium primum ASD (OP-ASD), but triphasic QRS pattern (rSR' or M pattern) in V1 is characteristic. rSR' pattern in V1 with a R' greater than S followed by inverted T wave which is commonly seen in volume overload Right Ventricular overloading/enlargement/hypertrophy (7). The right precordial leads shows QRS pattern like those found in OS-ASD. However, the left precordial leads may show sign of diastolic left ventricular overloading/hypertrophy/enlargement when marked mitral regurgitation or a large VSD, or both, are present.

- 1) P wave:** variable, normal, with LAE, RAE or BAE The mitral valve cleft causes significant mitral regurgitation, the left side of the heart also becomes volume overloaded. A left ventricle to right atrium shunt can be present, which further overloads both the right and left heart. Patients with mitral valve insufficiency may have left ventricular hypertrophy/enlargement or left atrial enlargement/overload.

- 2) **PR interval duration:** prolonged in 50% of the cases by increase of AV conduction time. First-degree heart block suggests an OP-ASD but may be seen in older patients with a OS-ASD.
- 3) **QRS axis (SÂQRS):** with extreme shift in superior quadrants and counterclockwise rotation (CCWR) of QRS loop in the frontal plane. When an OP-ASD is present, the ECG reveals extreme left axis deviation. When OS-ASD is present, the ECG reveals right axis deviation.
- 4) **QRS pattern: in II, III and aVF,** rS-type complexes with notch in the ascending limb of S wave. qR complexes in DI and aVL: LAFB-like pattern.
- 5) **aVR:** qR complexes with broader T wave;
- 6) **Ventricular overload:** More evident signs of right ventricular hypertrophy/enlargement (RVH/RVE) in its complete form (ostium atrium ventricularis comunis). There may be criteria for biventricular hypertrophy/enlargement (BVH/BVE) or diastolic, eccentric or volumetric left ventricular hypertrophy/enlargement (LVH/LVE). More evident signs of RVE in its complete form (ostium atrium ventricularis comunis). There may be criteria for BVH/BVE or LVH/LVE. Shunting is predominantly left-to-right in the absence of pulmonary vascular disease or significant right ventricular outflow tract obstruction. This results in volume overload of the right atrium and ventricle and pulmonary overcirculation;
- 7) **Triphasic QRS (rsR' or rSR')** nearly always present in right precordial leads V_{3R}, V₁ and V₂ and broader S wave in DI, aVL, V₅ and V₆.
- 8) **aVR: qR complexes with broader T wave;**
- 9) **Postoperative complications after repair**
 - AF or AFL may develop in approximately 5% of patients. Increased premature ventricular contractions are observed in up to 30% of patients. Complex ventricular arrhythmias occur most commonly when LV dysfunction is present. (8).
 - During the immediate postoperative period, persistent complete AV block occurs in 1% to 7% of patients and 2% of patients thereafter (9). Prolonged infra-Hisian conduction time may indicate increased risk of late AV block, even in the presence of a previous normal PR interval (10).

The Vectorcardiogram in OP-ASD

In patients with OP-ASD (patients with only atrial septal defect) right ventricular systolic pressure is less than 70 mm Hg. Patients with right ventricular systolic pressure greater than 70 mm Hg are made up predominantly of patients with complete AV canals.

The normal value of Maximal Spatial Voltage to the Right (RMSV) is 0.6 mV in average, this parameter was found to relate well to the level of right ventricular systolic pressure. Thus, Right ventricular systolic pressure less than 40 mm Hg the mean RMSV is only 1.2 mV. All patients with an RMSV less than 1 mV have a normal right ventricular systolic pressure

Right ventricular systolic pressure greater than 100 mm Hg RMSV = 3.3 mV in average.

Patient with RMSV greater than 1.7 mV occur in patient with right ventricular systolic pressure over 40 mm Hg (11).

P-loop: is directed to the left and inferiorly with normal or increase magnitude. In presence of significant mitral regurgitation(cleft) the left atrium dilates and produce increase posterior and inferior forces.

QRS-loop

Horizontal Plane

The QRS rotation of QRS loop reflect primarily the right systolic pressure, thus, counterclockwise rotation is observed in OP-ASD with minor right ventricular systolic pressure (less than 40 mm Hg). Figure in-eight or clockwise QRS-loop rotation is observed predominately in complete AV canals with major right ventricular systolic pressure.(major than 70 mm Hg). A left posterior orientation of QRS loop may be seen with marked mitral regurgitation (cleft) and/or a large ventricular septal defect in association to OP-ASD.

Frontal Plane

In approximately 95% of cases characteristic initial forces directed inferiorly, the QRS loop is has counterclockwise rotation with superior frontal QRS-loop (extreme left axis deviation) very similar pattern with truly left anterior fascicular block. In few cases QRS loop has narrow figure-eight QRS-loops in a linear figure-eight manner that is slightly superior to the 0° to 180 axis and tends to be parallel to this axis. In these cases the maximal QRS vector is also directed to the left and superiorly.

When QRS duration is within normal limits for age, the frontal QRS-loop became superior by 30 ms. On the other hand in cases with QRS duration prolonged QRS loop became superior at a mean time of 40 ms.

The principal difference between OS-ASD and OP-ASD (endocardial cushion defects) consists in the loop being inferior in the former, superior in the latter. The direction of inscription in the frontal plane tends to be clockwise in the OS-ASD counterclockwise in the endocardial cushion group (12; 13).

Right Sagittal Plane: counterclockwise superior rotation.

T-loop: oriented to the left and inferiorly, which is discordant to the QRS loop in the frontal plane.

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