FALLOT'S TETRALOGY(TOF) ELECTROVECTORCARDIOGRAPHIC FEATURES BEFORE AND AFTER TOTAL CORRECTION NON-INVASIVE, PROCEDURES FOR SUDDEN CARDIAC DEATH PREVENTION

Synonymous:
Fallot's syndrome
Fallot's tetrad
Steno-Fallot tetralogy
La Maladie blue
Blue baby syndrome

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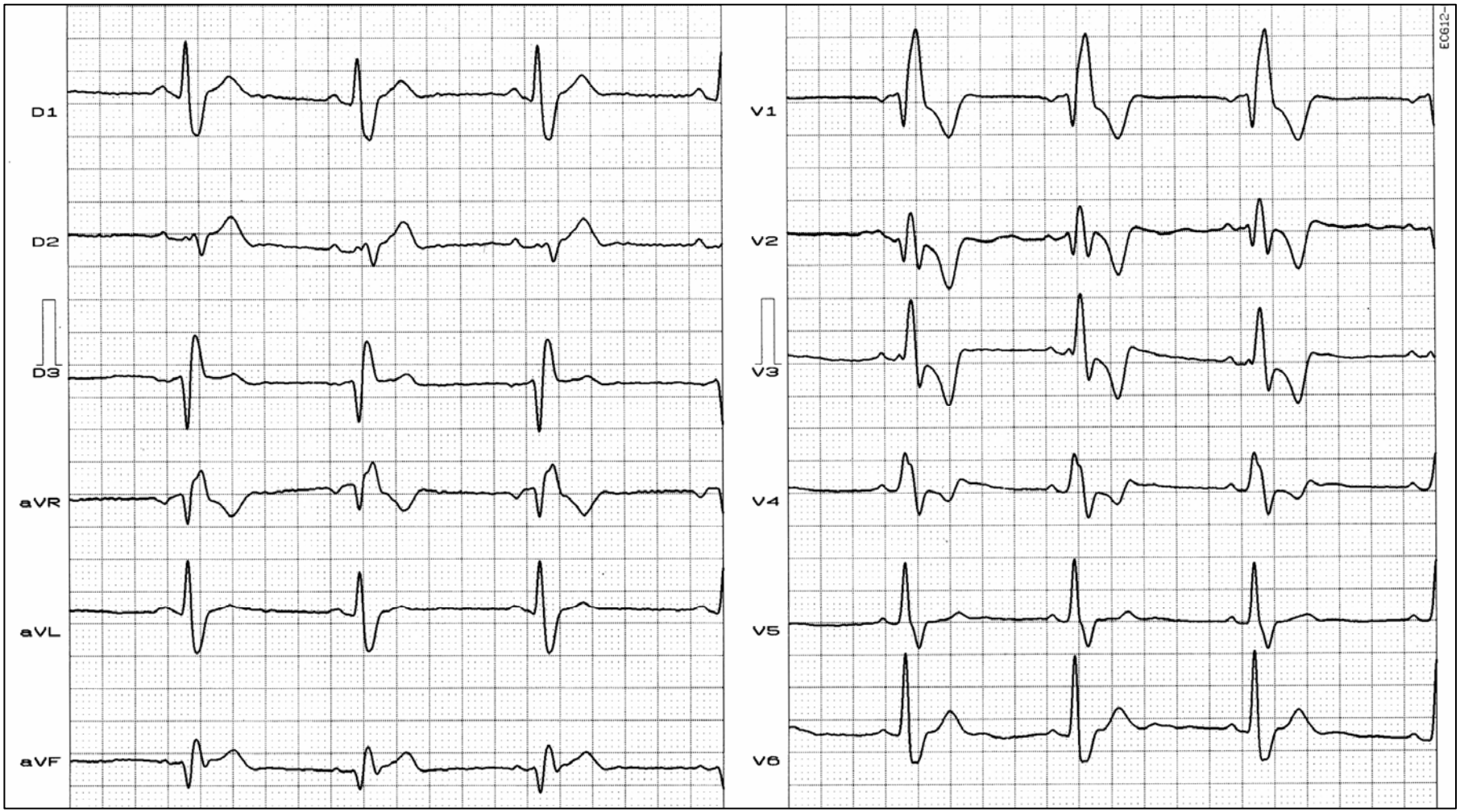
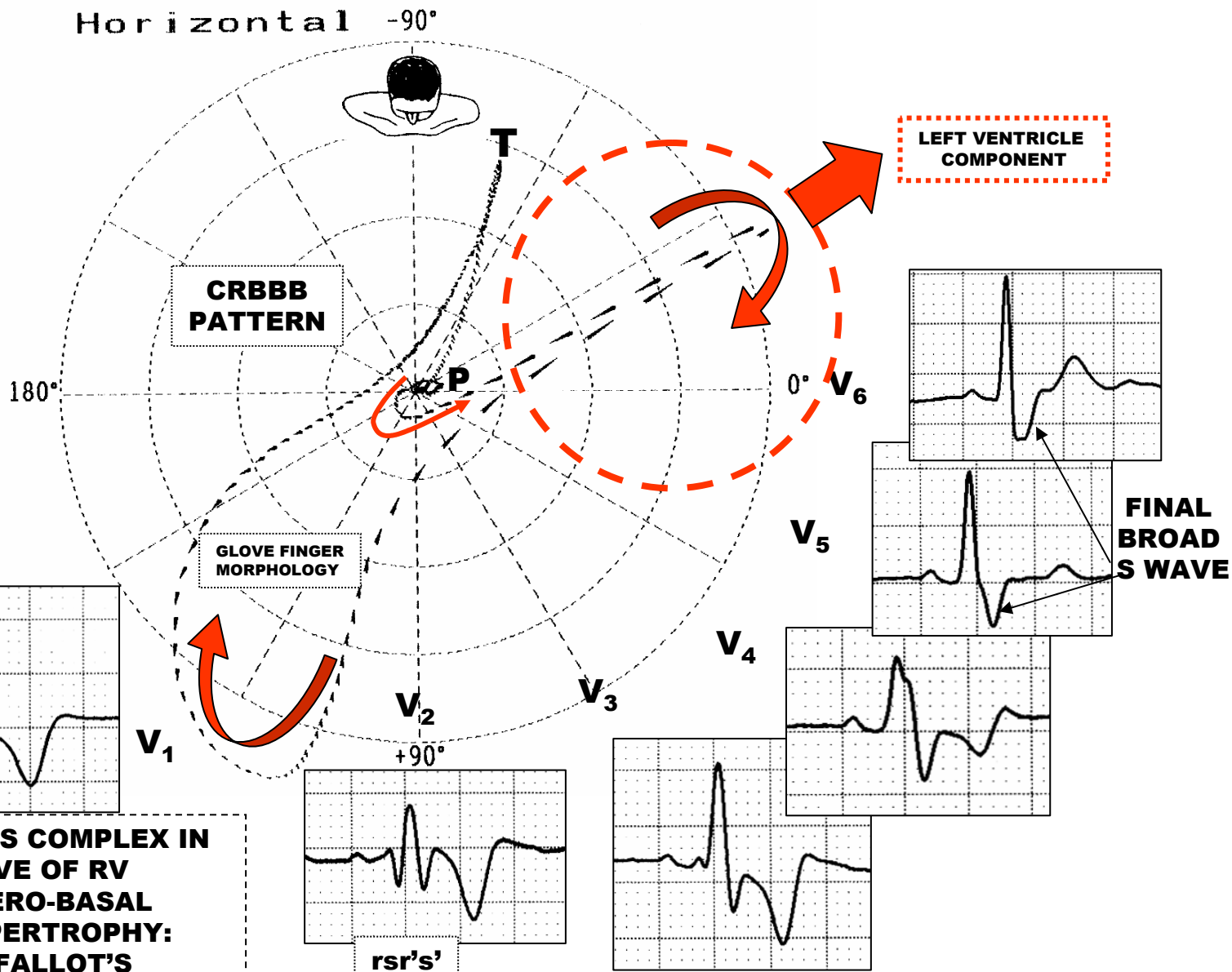
Name: FV ; **Gender:** Male; **Age:** 26 years old; **Ethnic group:** Caucasian; **Weight:** 77 Kg;
Height:1,80 m **Biotype:** Normoline **Date:** 25/09/2007



Clinical Diagnosis: late postoperative (25 years ago) of tetralogy of Fallot repair(total correction). The surgeon close the ventricular septal defect with a patch and opens the right ventricular outflow tract by removing some thickened muscle.

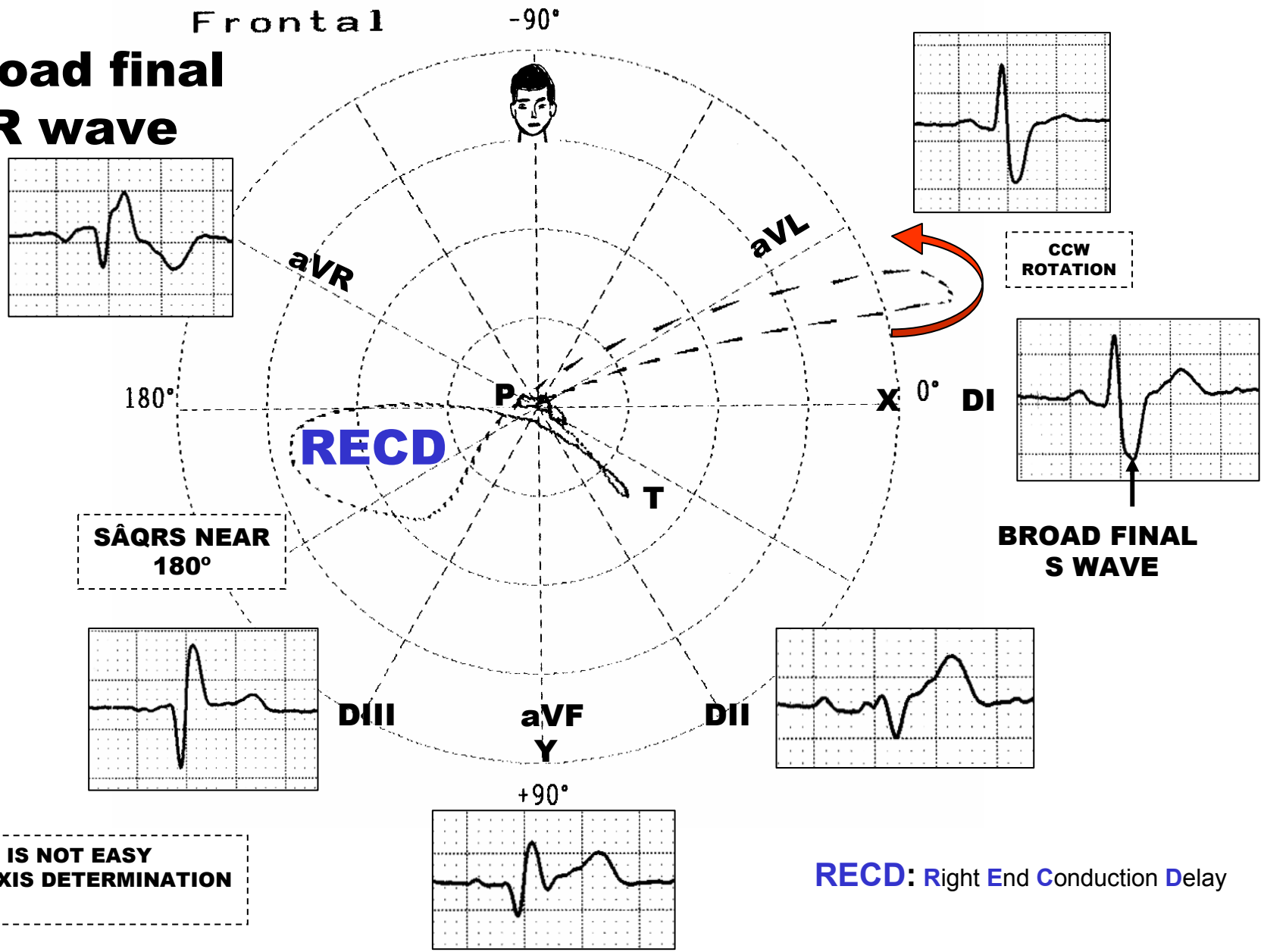
ECG diagnosis: QRSd: 190ms, broad tetraphasic complex in V2 indicative of RV lateroposterobasal selective hypertrophy, fragmented wide QRS (f-wQRS), S final wave on left leads I, aVL, V5 and V6, prominent final R wave in aVR: Complete Right Bundle Branch Block. Typical of postoperative ECG pattern Fallot's Tetralogy.

ECG/VCG CORRELATION HORIZONTAL PLANE

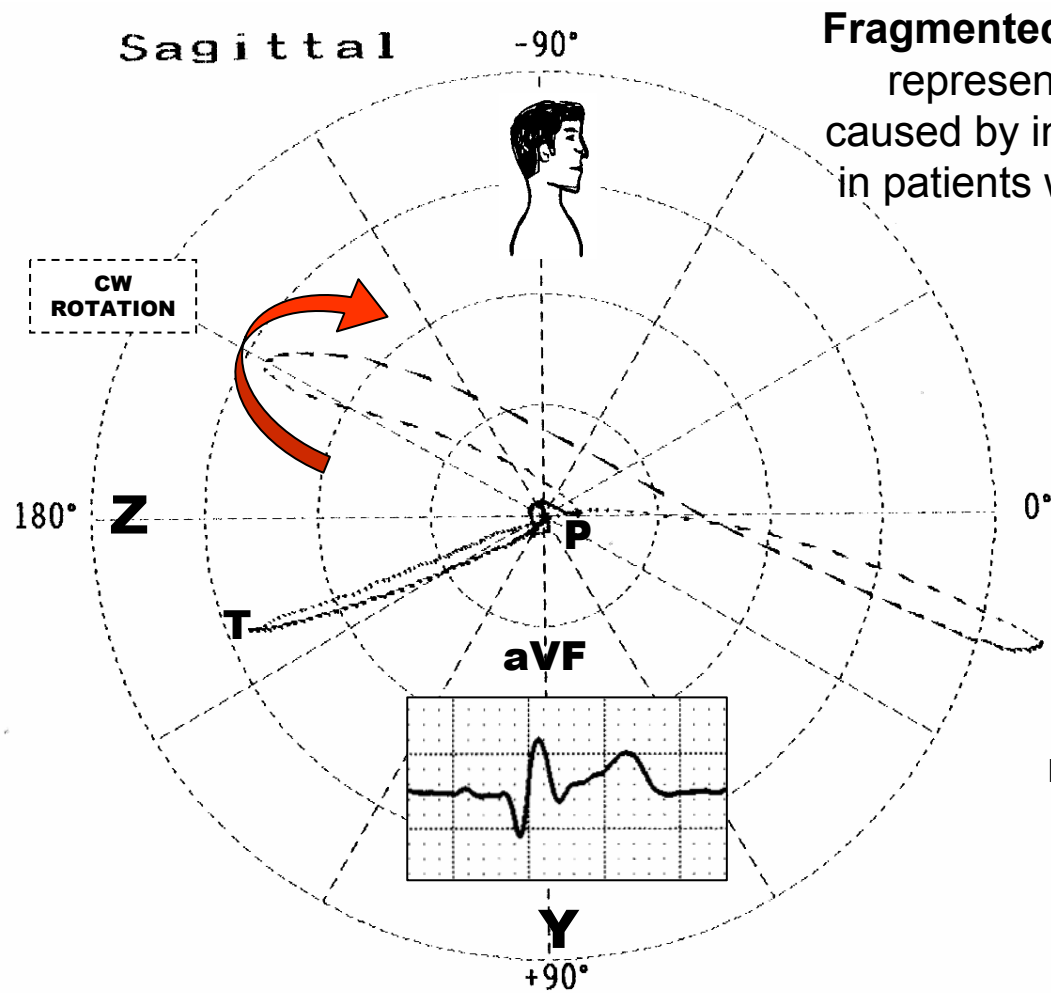


TETRAPHASIC QRS COMPLEX IN V2 INDICATIVE OF RV LATEROPOSTERO-BASAL SELECTIVE HYPERTROPHY: TYPICAL OF FALLOT'S TETRALOGY. FRAGMENTED WIDE QRS (f-wQRS)

ECG/VCG CORRELATION FRONTAL PLANE

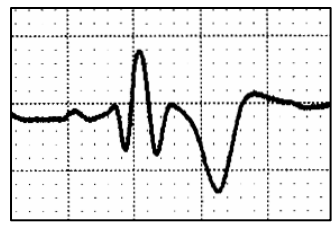


ECG/VCG CORRELATION RIGHT SAGITTAL PLANE



Fragmented wide QRS (f-wQRS): represents conduction delay caused by incision myocardial scar in patients with T4F after repair([1])

rsr´s´

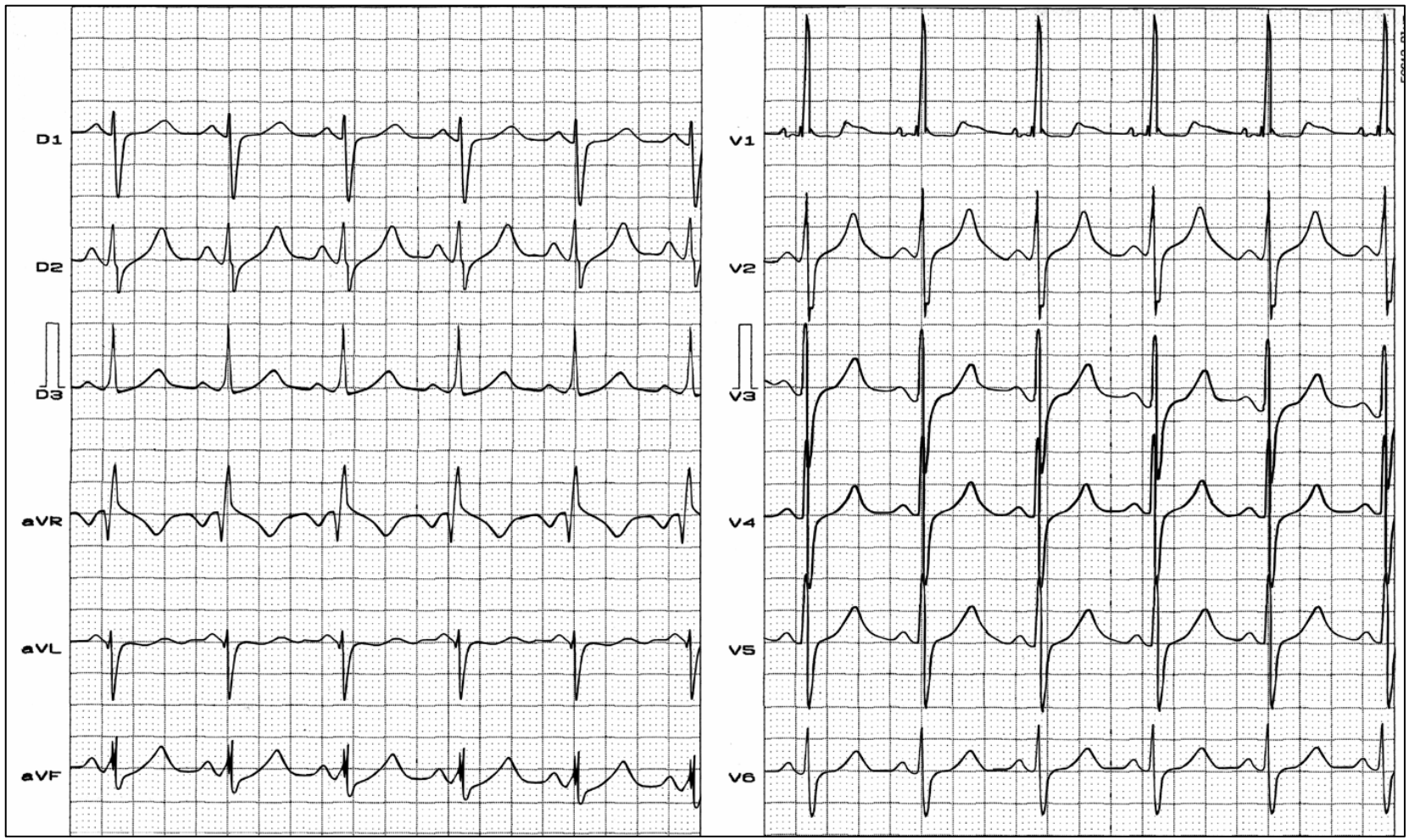


TETRAPHASIC QRS COMPLEX IN V2 INDICATIVE OF RV LATEROPOSTERO-BASAL SELECTIVE HYPERTROPHY: TYPICAL OF FALLOT'S TETRALOGY

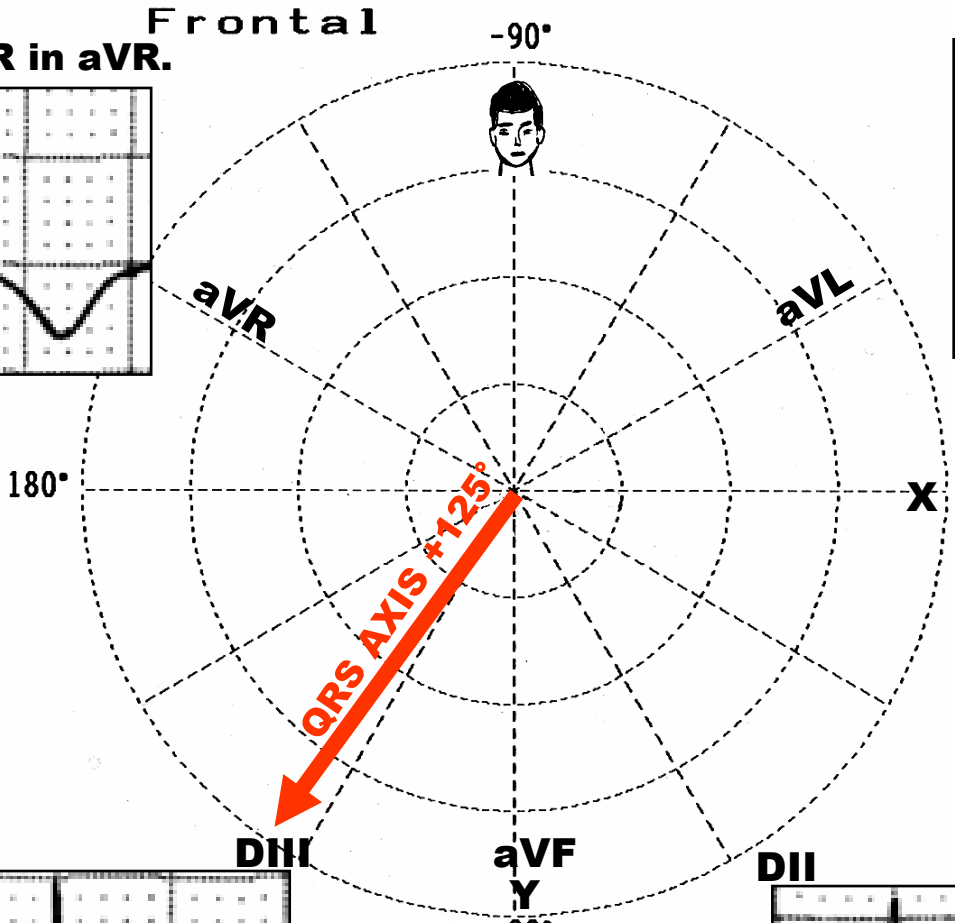
1. Jacob S, Agarwal K, Afonso L. QRS fragmentation in patients with repaired tetralogy of Fallot. Am J Cardiol. 2009 Sep 1;104:740-741.

ECG IMMEDIATELY BEFORE THE SURGERY

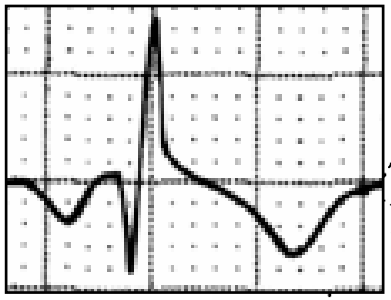
Name: FV ; **Gender:** Male; **Age:** 20 days; **Ethnic group:** Caucasian; **Weight:** 2,900 gr.
Height: 47 cm; **Date:** 04/09/1981

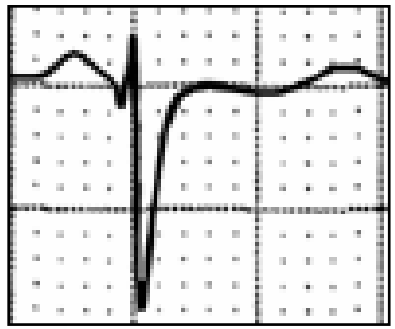


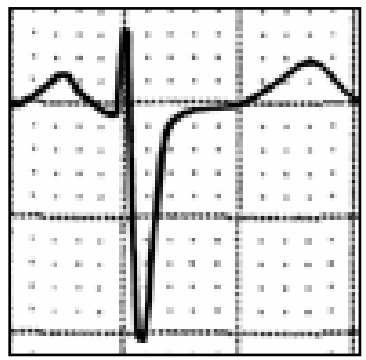
ECG/VCG CORRELATION FRONTAL PLANE



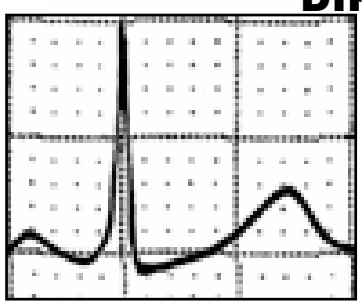
Prominent R in aVR.

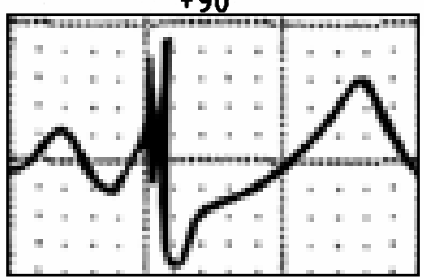


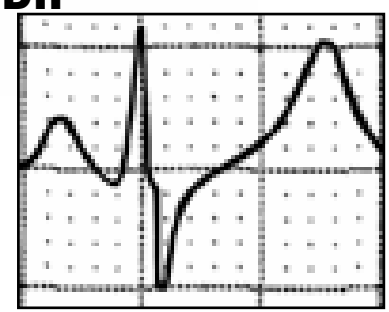




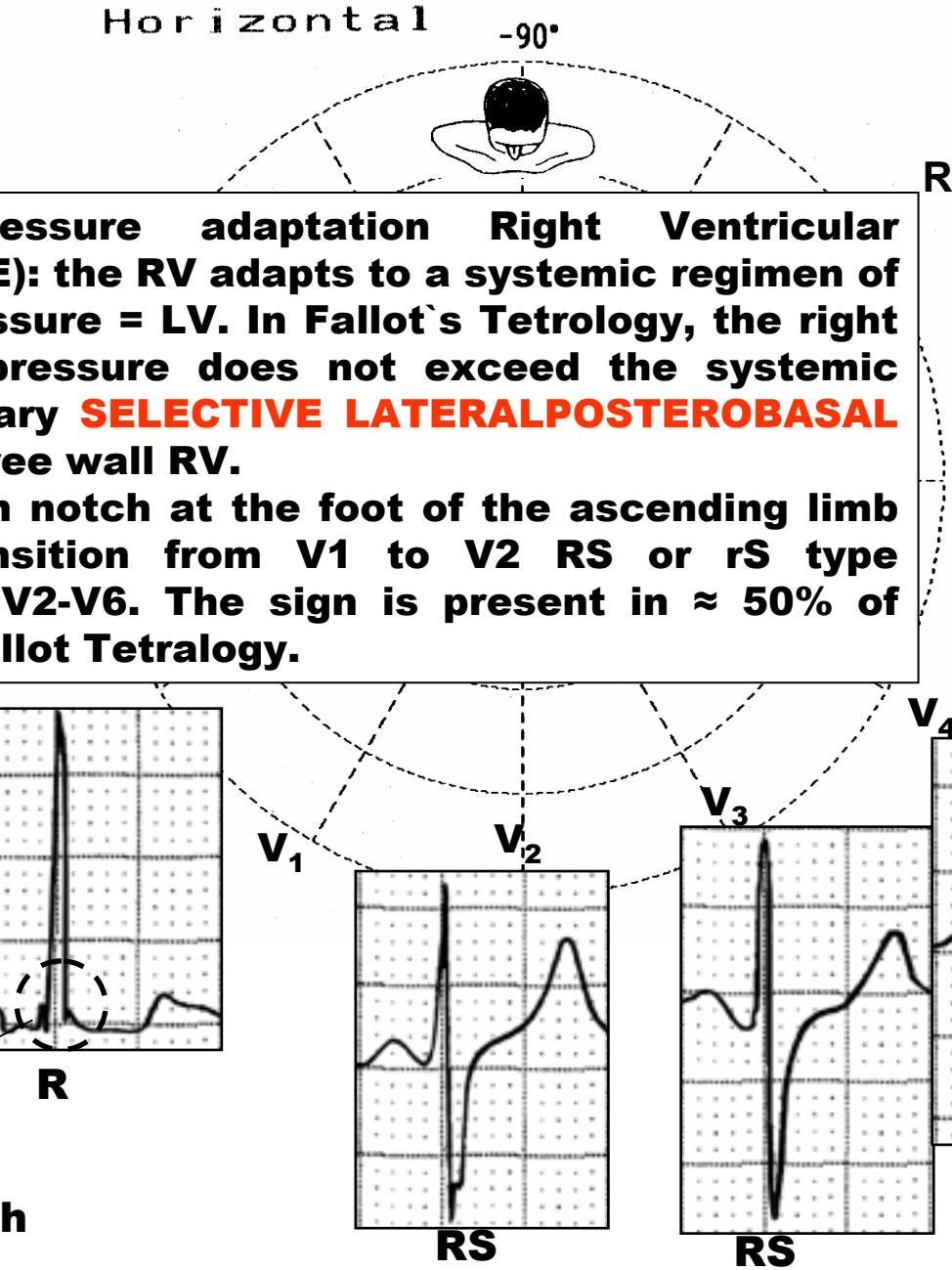
In normal newborn babies, average QRS axis is around +125°, and it may reach 180°, i.e. in the right inferior quadrant.







ECG/VCG CORRELATION HORIZONTAL PLANE



Systolic or pressure adaptation Right Ventricular Enlargement (RVE): the RV adapts to a systemic regimen of pressure: RV pressure = LV. In Fallot`s Tetrology, the right intraventricular pressure does not exceed the systemic one with secondary SELECTIVE LATERALPOSTEROBASAL GROWTH of the free wall RV.

Pure R wave with notch at the foot of the ascending limb and sudden transition from V1 to V2 RS or rS type complexes from V2-V6. The sign is present in ≈ 50% of these cases of Fallot Tetralogy.

R/S ratio in V5 and V6 ≥ 1.

R with initial notch

MAIN ECG FEATURES IN TOF BEFORE CORRECTION

P wave: Right Atrial Enlargement (RAE) is rare in childhood. Exceptional before 1 year old. RAE is frequent in older children(1: 4). 65% of adult show RAE.

QRS axis: moderate to marked right-axis deviation is the rule(+120° to 210°) is the rule, except in termed "atypical group": T4F with increased pulmonary flow, associated to Noonan syndrome, associated to ostium primum atrial septal defect.

QRS complex: Abnormal Q/R ratio in aVR in >60% of cases: Prominent final R wave > 5 mm (RVOT). Positive QRS complexes in V3R and V1: R, Rs or R with notch on initial portion of ascending R wave. Rs with RS ratio > 1. R wave of V1 ≥ 7 mm, isodiphasic or negative QRS complexes from V2 to V6 (RS or rS) This phenomena is observed in approximately 50% of cases and is indicative that the right intraventricular pressure does not exceed the systemic one([1]). Selective lateral-posterior-basal growth of the RV, regression of R/S ratio in the precordium, incomplete RBBB (in 20% of cases), complete RBBB (common in adults).

Biventricular ventricular enlargement is observed in cases with increased pulmonary flow([2]).

T wave: T vector may have a normal orientation: to the left, below and to the front. Positive T wave in V1 after 3 days of life and up to 6 years of age, when the R/S ratio in this lead is greater than 1 is indicative of RVH([3; 4]).

1. Pileggi F, Bocanegra J, Tranchesi J, et al. The electrocardiogram in tetralogy of Fallot: a study of 142 cases. Am Heart J. 1960 May; 59: 667-680.
2. Astrakhantseva GI, Bukharin VA. [2 cases of tetralogy of Fallot with a left type of the electrocardiogramGrudn Khir. 1961 Mar-Apr;3:101-104.
3. Hansoti RC, Shah VV, Anklesaria CM. The electrocardiogram in the tetralogy of Fallot. A study of seventeen cases. Indian Heart J. 1965 Jul;17:229-233.
4. Hansoti RC, Shah VC, Anklesaria CM. The Electrocardiogram In The Tetralogy Of Fallot. A Study Of Seventeen Cases. Indian Heart J. 1965 Jan;17:34-39.

MAIN VCG FEATURES IN TOF

•Initial 20ms vector preserved and directed to the front an rightward([1]).

•Type A or B right ventricular enlargement pattern Quantitative analysis demonstrated the constant increase of the RMSV and the direct relationship of the LMSV to arterial oxygen saturation, and so it was the principal parameter for evaluation of the left ventricular volume and the size of pulmonary flow and the degree of pulmonary stenosis([2]).

• QRS loop in the horizontal plane (H) keeps a clockwise(CW), figure in eight or counterclockwise(CCW) rotation. Qualitative analysis showed a CW or figure eight QRS loop on the H and F planes in the great majority of cases; and only in a CCW loop on the H plane is the suspicion of an arterial oxygen saturation greater than 85% especially in cases older than two months.

•>70% of the area of the QRS loop on horizontal plane located on anterior quadrants([3]).

•The terminal forces of the QRS loop on the H plane is always directed under the O point, whereas in the valvular pulmonary stenosis, terminal forces are generally superior to the O point.

•The relationship between the azimuth of 0.01 sec spatial vector to O2 saturation is highly significant, showing a progressive anterior development of the former, as the latter increased([4]).

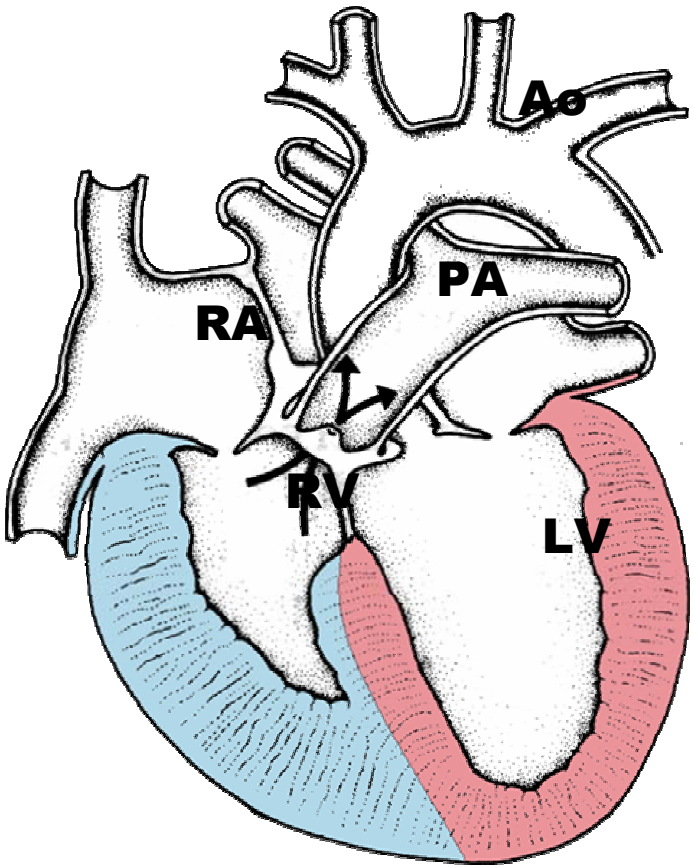
1. **Depasquale NP, Burch GE. The electrocardiogram, vectorcardiogram, and ventricular gradient in the tetralogy of Fallot. Circulation. 1961 Jul;24:94-109.**
2. **Marsico F, Calabrò R, Alborino A, Mininni N. The vectorcardiogram of Fallot's tetralogy in the first two years of life. Qualitative and quantitative analysis G Ital Cardiol. 1976;6(6):1070-81.**
3. **Makolkin VI, Zargarli FI, Namazova AA, Belyaeva AI, Korotkov AA. Electrocardiogram and vectorcardiogram in the Fallot tetralogy.Cor Vasa. 1967;9:191-199.**
4. **Aihara N.Vectorcardiographic studies on Fallot syndrome, with special reference to the marious types of vectorcardiographic patterns and the degrees of the clinical severityJpn Circ J. 1968 Jun;32:899-908.**

HEMODYNAMIC MODALITIES OF RVE

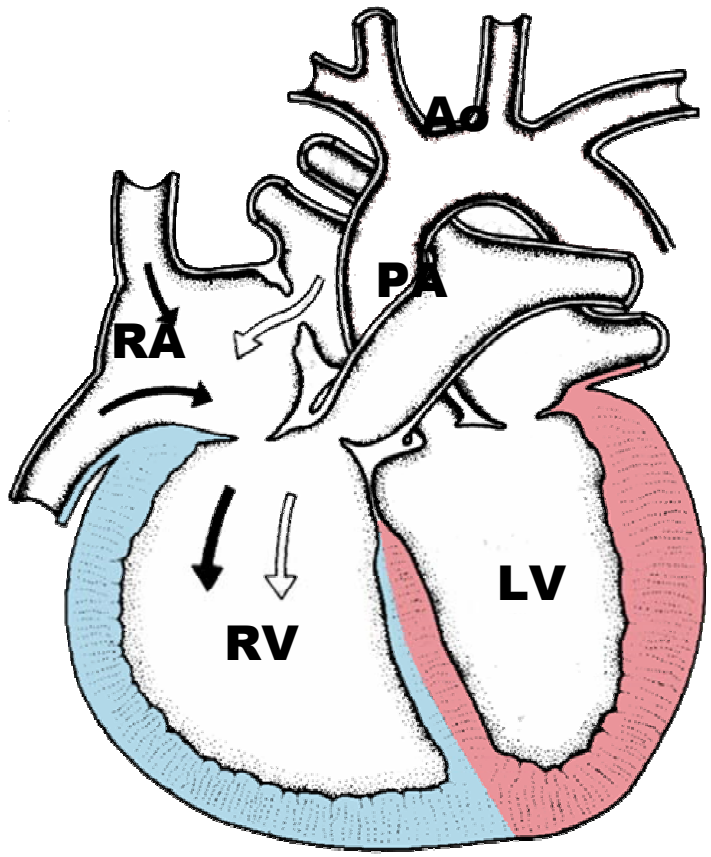
I) Systolic or pressure RVH:

Ia) Systolic of adaptation (our case)

Ib) Systolic with strain pattern



II) Diastolic, Eccentric or Volumetric RVH or RVE



HEMODYNAMIC MODALITIES OF RVE

I) Systolic or Pressure RVH or RVE

Ia) Systolic of adaptation (our case)

- ✓The RV adapts to a regimen of systemic pressure
- ✓The right intraventricular pressure does not exceed the systemic one
- ✓Selective lateral-posterior-basal growth of the free wall right ventricle
- ✓QRS morphologies in V1 and V3R may be: R, RS or R with initial notch (our case).
- ✓V2 and V3 show RS or rS pattern because they face the inferior region of the free RV wall and trabecular area that present little hypertrophy.

Ib) Systolic with strain pattern (severe RVE)

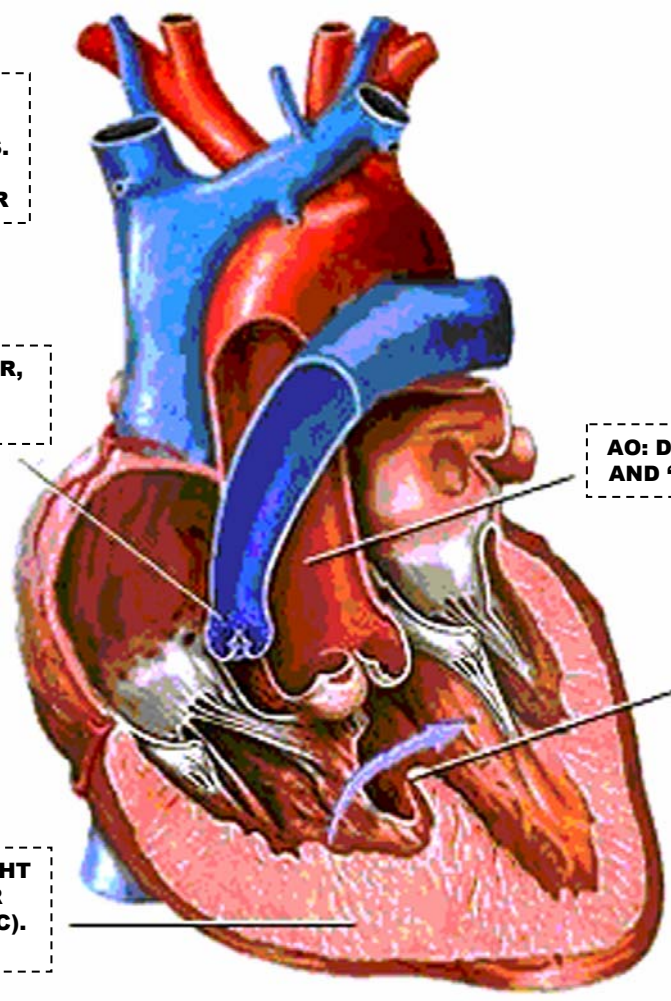
- ✓Global RVE
- ✓Right ventricular intraventricular pressure may exceed the systemic one
- ✓V2 and V3 leads continue showing QRS of positive predominance
- ✓Inverted T wave with a tendency to be symmetrical (primary) on right precordial leads.
- ✓Typical example: severe pulmonary stenosis.

II) Diastolic, Eccentric or Volumetric RVH or RVE

- ✓RBBB pattern: triphasic morphology of QRS complexes in V3R, V1 and V2 with <120 ms duration: IRBBB, CRBBB or End Conduction Delay
- ✓Final broad S wave in the left leads I, aVL, V5 and V6
- ✓Final prominent broad R wave of aVR

TETRALOGY OF FALLOT (TOF)

PREVALENCE:
400 CASES EACH 100,000 BABIES BORN
9 TO 10% OF CONGENITAL HEART DISEASES.
THE MOST FREQUENT ONE IS CYANOTIC CONGENITAL HEART DISEASE AFTER 1 YEAR



PA: HYPOPLASTIC WITH VALVULAR, INFUNDIBULAR OR INFUNDIBULO-VALVAR PS.

AO: DEXTROPOSITIONED, LARGE AND "OVERRIDING" THE SEPTUM

VSD: MID-BASAL, INFRACRITSAL AND LARGE (GREATER THAN 1 cm^2/m^2).

CYANOSIS

RVE: SECONDARY (RIGHT INTRAVENTRICULAR PRESSURE = SYSTEMIC). HYPERTROPHIC RV

Prevalence and anatomical features of T4F.

FALLOT´S TETRALOGY DISCOVERY

Was first described in 1671 by the Danish physician, natural scientist, Bishop – and later was described in 1672 by geologist and anatomist Saint Niels *Steensen or* Stensen (Nicolaus Stenonius). In 1773 by Edward Sandifort, and in 1888 by the French physician Etienne-Louis Arthur Fallot (1850-1911)([1]). The last one was a French physician renowned for his painstaking physical examination.

At least some forty case descriptions precluded that of Fallot. His delineation from 1888 was based on details of the pathological features of two persons with the condition, together with a survey of some 50 previous observations. Fallot considered the deformity as an entity and as such the most common cause of cardiac cyanosis. By his contemporaries Fallot's observations were considered to be solely of theoretical or curious interest. Congenital heart malformations attracted little attention, as there was still no therapy available.

Fallot's work first received serious attention when Paul Dudley White (1886-1973) in 1931 discussed the malformation and translated Fallot's works.

1. **Fallot ELA:** ***Contribution à l'anatomie pathologique de la maladie bleue (cyanose cardiaque).*** **Marseille médical, 1888, 25: 77-93, 138-158, 207-223, 341-354, 370-386, 403-420.**

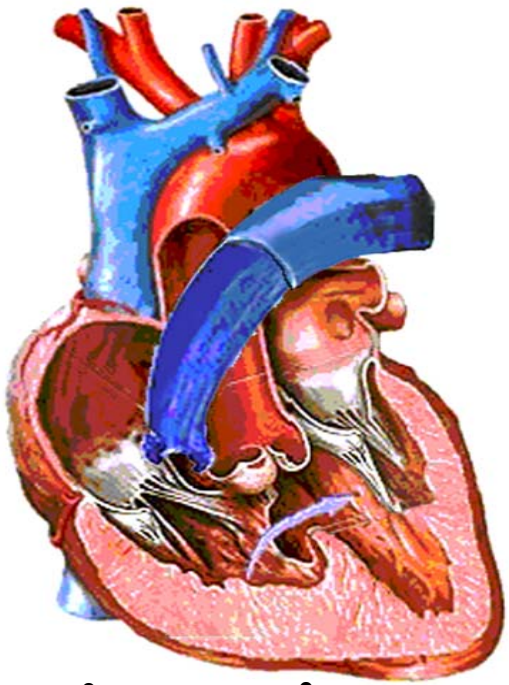
FALLOT´S TETRALOGY: ANATOMY

There are four characteristic abnormalities in tetralogy of Fallot (TOF):

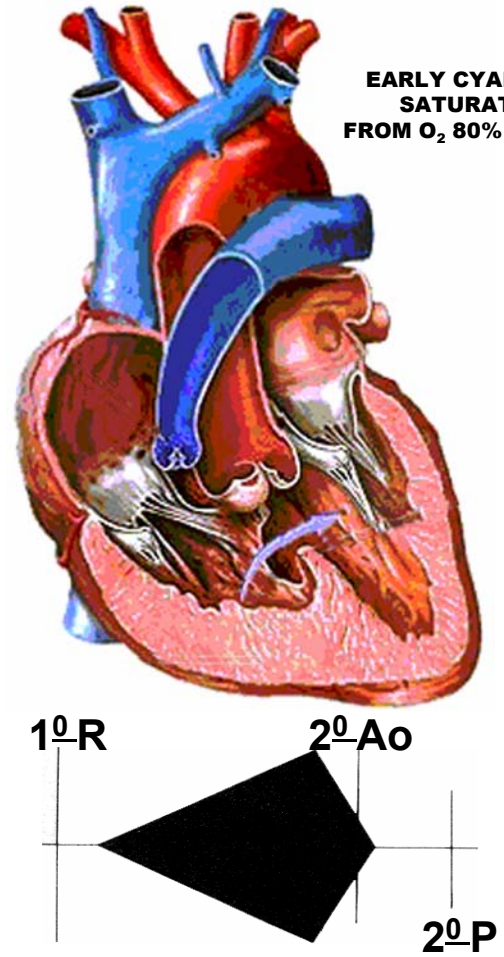
1. **Right Ventricular Outflow Tract (RVOT) obstruction: hypoplastic with valvular, infundibular or infundibulo-valvar pulmonary stenosis**
2. **Ventricular Septal Defect(VSD): mid-basal, infracritsal and usually large (greater than 1 cm2/m2) just below the aortic root**
3. **Aortic root: dextropositioned, large and "overriding" a VSD There is also right-sided aortic arch in around 20% cases**
4. **Right Ventricular Enlargement/Hypertrophy (RVE/RVH): secondary (right intraventricular pressure = systemic). Right Ventricle Hypertrophy.**

Atrial Septal Defect (ASD) in 8-10% Pentalogy of Fallot.

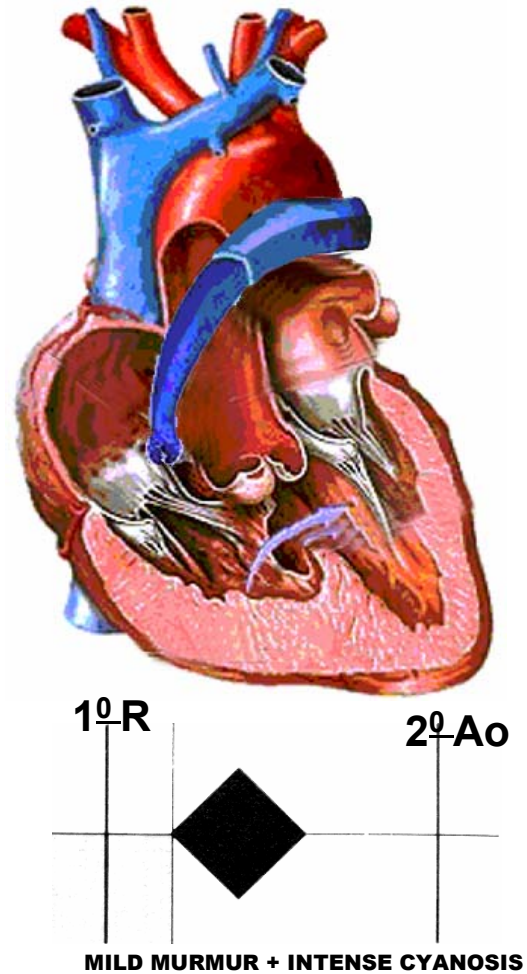
PINK-FALLOT	CLASSICAL T4F (MODERATE)	T4F WITH PULMONARY ATRESIA, PSEUDO-TRUNCUS OR EXTREME



Intense murmur + mild cyanosis
Less evident RVE
Late or absent cyanosis
Saturation from O_2 90% to 100%.



Systolic RVE of adaptation: R/S (R>S) in V3R and V1 with notch at the foot of the R wave and sudden transition from V1 to V2, i.e. predominantly positive QRS complexes in V1 for rS type complexes in V2. The sign is present in 50% of these cases.



The three types of T4F (mild, moderate and extreme). Characteristics of the murmur.

Late Complications of Repair of Tetralogy of Fallot And Indications For Pulmonary Valve Replacement([1]).

1.Chronic Pulmonary Regurgitation

2. **Right Ventricular Dilation:** Severe right ventricular dilatation and pulmonary regurgitation secondary to outflow tract repair in TOF are frequently occurring sequelae developing slowly over time. Indications for pulmonary valve replacement remain controversial because echocardiographic findings or arrhythmias are not always accompanied by deterioration of clinical condition. However, right atrial dilatation and additional hemodynamic lesions demand increased vigilance. Transatrial repair is associated with favorable outcome **([2])**

3. Electrical Inhomogeneity

4. Myocardial Scarring

5. Wenckebach type block secondary to infra-Hisian location ([3]).

Complete repair in patients with complete atrioventricular septal defect associated with the TOF seems to offer acceptable early and mid-term outcome in terms of mortality, morbidity, and reoperation rate. Palliation prior to complete repair may be reserved in specific cases presenting small pulmonary arteries or severely cyanotic neonates. The RVOT should be managed in the same fashion as for isolated TOF; however, a transatrial transpulmonary approach is approach of choice **([3]).**

1) **Cheung MM, Konstantinov IE, Redington AN. Late complications of repair of tetralogy of Fallot and indications for pulmonary valve replacement.Semin Thorac Cardiovasc Surg. 2005 Summer;17(2):155-159.**
2) **de Ruijter FT, Weenink I, Hitchcock FJ, Meijboom EJ, Bennink GB.; discussion 1800.Right ventricular dysfunction and pulmonary valve replacement after correction of tetralogy of Fallot. Ann Thorac Surg. 2002 Jun;73(6):1794-800.**
3) **Marijon E, Combes N, Boveda S, Albenque JP. Wenckebach type block on surface ECG due to infra-Hisian location in a patient with repaired tetralogy of Fallot. Wenckebach type block on surface ECG due to infra-Hisian location in a patient with repaired tetralogy of Fallot.**
4) **Prifti E,Bonacchi M, Bernabei M, Leacche M, Bartolozzi F, Murzi B, Battaglia F, Nadia NS, Vanini V._Repair of complete atrioventricular septal defect with tetralogy of fallot: our experience and literature review. J Card Surg. 2004 Mar-Apr;19:175-183.**

In a retrospective study, 99 adult patients with TOF, who had undergone a first pulmonary valve replacement late after initial total correction, were identified. Computer-generated QRS durations (QRSd) were obtained from ECG.

A mixed linear regression model was used to analyze the course of QRSd over time and to identify risk factors for increase in QRSd over time. Composite end point was created from SCD, VT or ICD discharge.

In total, 99 patients (57% men, mean (SD) age at pulmonary valve replacement 29 (11) years) with a median follow-up of 4.9 (0.1-16) years were analyzed.

In patients with preoperative QRS <120 ms, surgery caused no significant change in QRSd, and after surgery, QRSd remained stable over time. By contrast, in patients with a preoperative QRS of 150-180 ms or QRS ≥180 ms, surgery resulted in QRS shortening. During follow-up, a QRS widening 1.1(1.3) ms/year ($p<0.001$) in both groups was observed. In patients with a preoperative QRSd ≥180 ms, no significant difference was observed in the number of patients reaching the composite end point compared with patients with a preoperative QRSd of 150-180 ms. However, the former more often reached QRSd ≥180 ms again after surgery compared with the latter). None of the patients with a preoperative QRSd ≥180 ms died during follow-up. The authors observed a decrease in QRSd directly after surgery, followed by a steady increase, in patients with a preoperative QRSd >150 ms. The beneficial effect of pulmonary valve replacement on QRSd was transient. The risk of developing ventricular arrhythmias after surgery was substantial when preoperative QRSd was ≥180 ms, but mortality remained low.

1. **Oosterhof T, Vliegen HW, Meijboom FJ, Zwinderman AH, Bouma B, Mulder BJ. Long-term effect of pulmonary valve replacement on QRS duration in patients with corrected tetralogy of Fallot. Heart. 2007 Apr;93:506-509.**

Signal-Averaged Electrocardiogram (SAECG) value in determining ventricular arrhythmia risk in patients after Fallot repair

Early detection of arrhythmias after congenital heart disease surgery is important because it can help decrease morbidity and mortality. ECGs contain frequencies between 0.05 and 100 Hz, but higher frequencies are also present. Using SAECG, the highest amplitudes of these high-frequency components within the QRS complex can be recorded and analyzed. Omeroglu et al **([1]).** studied the relationship between ventricular late potentials (LPs), ventricular arrhythmias and right ventricular systolic pressure in 22 patients who underwent TOF repair (mean follow-up, 40.1 +/- 33.5 months). Holter ECG monitoring and SAECGs were performed. SAECG parameters studied included the duration of the filtered QRS, the duration of terminal QRS below 40 muV, and the root mean square amplitude of the terminal 40ms. Cardiac catheterization was performed on 19 patients. 18 healthy volunteers were studied as a control. Ventricular arrhythmias were found in 13 patients; right ventricular systolic hypertension was found in 1 patient. No significant residual ventricular septal defects were detected. 8 patients had ventricular LPs. Right ventricular systolic pressure did not differ significantly between patients with or without LPs. There were significant differences between patients with ventricular arrhythmias and healthy volunteers; filtered QRS duration was significantly longer in patients with ventricular arrhythmias. SAECG may be beneficial in determining ventricular arrhythmia risk in tetralogy of Fallot patients postoperatively.

1. **Omeroglu RE, Olgar S, NisliK Signal-averaged electrocardiogram may be a beneficial prognostic procedure in the postoperative follow-up tetralogy of fallot patients to determine the risk of ventricular arrhythmias..Pediatr Cardiol. 2007 May-Jun;28: 208-212.**

Fragmented QRS (fQRS) and Fragmented wide QRS (f-wQRS)

DEFINITIONS

I) Fragmented QRS (fQRS)

It is defined as conduction abnormalities (depolarization abnormality) not typical of a bundle branch block, within the QRS complex with secondary changes in QRS morphology, QRS duration ≤120ms, with notches or slurs of QRS complexes with different RSR´ patterns: additional R waves, notched S wave, or > 1 R´ wave ([1]). When studied in coronary artery disease it is necessary to be present at least on two contiguous leads.

II) Fragmented wide QRS (f-wQRS):

It is the same with fQRS duration≥120ms**)([2]).**

1. **Pietrasik G, Goldenberg I, Zdzienicka J, Moss AJ, Zareba W. Prognostic significance of fragmented QRS complex for predicting the risk of recurrent cardiac events in patients with Q-wave myocardial infarction. Am J Cardiol. 2007 Aug 15; 100: 583-586.**
2. **Das MK, Suradi H, Maskoun W, Michael MA, Shen C, Peng J, Dandamudi G, Mahenthiran J. Fragmented wide QRS on a 12-lead ECG: a sign of myocardial scar and poor prognosis. Circ Arrhythm Electrophysiol. 2008 Cot: 1:258-268.**

The fQRS complex on ECG is another marker of depolarization abnormality.

fQRS is associated with increased mortality and arrhythmic events in patients with:

1. Coronary Artery Disease: fQRS on ECG is associated with myocardial scar in patients with CAD. The presence of fQRS independently of Q waves are not associated with increased risk of recurrent events in the general population of patients after MI. However, among patients with resolved Q waves, fQRS is associated with increased risk of cardiac events **([1]).**

f-wQRS (fQRS with duration≥120ms) on ECG is a moderately sensitive and highly specific sign for myocardial scar in patients with known or suspected coronary artery disease. f-wQRS is also an independent preditor of mortality **([2]).** The fQRS is an independent predictor of cardiac events in patients with CAD. It is associated with significantly lower event-free survival for a cardiac event on long-term folow-up([3]). fQRS complexes on an ECG are a marker of higher stress MPI perfusion and functional abnormalities. Regional FQRS patterns denote the presence of a greater corresponding focal regional myocardial scar on stress myocardial perfusion imaging **([4]).** The fQRS on ECG is a marker of a prior MI, defined by regional perfusion abnormalities, which has a substantially higher sensitivity ad negative predictive value compared with the Q wave. This regress or even disappear over time, and there is no specific ECG sign of a non-Q wave MI([5]).

1. **Pietrasik G, Goldenberg I, Zdzienicka J, Moss AJ, Zareba W. Prognostic significance of fragmented QRS complex for predicting the risk of recurrent cardiac events in patients with Q-wave myocardial infarction. Am J Cardiol. 2007 Aug 15; 100: 583-586.**
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1. **Nonischemic dilated cardiomyopathy with a narrow QRS interval and sinus rhythm:** fQRS in the resting ECG is associated with significant intraventricular dyssynchrony in patients with nonischemic cardiomyopathy, narrow QRS and sinus rhythm. fQRS in ECG might be useful in identifying patients who could benefit from cardiac resynchronization therapy **([1]).**

2. **Arrhythmogenic right ventricular dysplasia:** fQRS in ARVD/C has a high diagnostic value similar to epsilon potentials by a highly amplified and modified recording technique **([2]).**

3. **Cardiac sarcoidosis:** The sensitivity and specificity of fQRS for detecting abnormal Gadolinium-delayed enhancement images were 100% and 80%, respectively. Sensitivity and specificity of Q waves were 11% and 100%, respectively. fQRS on ECG in patients with suspected cardiac sarcoidosis are associated with cardiac involvement as detected by Gadolinium-delayed enhancement on cardiac magnetic resonance imaging **([3]).**

4. **Fallot Tetralogy ([4])** and others congenital heart disease.

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5. **Brugada syndrome:** the presence of fQRS predicts episodes of VF during follow-up. epicardial activation delay led to fQRS, a risk marker of prognosis in BrS. Body surface mapping in patients with BrS supported these experimental findings. The AP heterogeneity within the epicardium of the RVOT contributes to the ECG characteristics, temperature sensitivity, TWA, and arrhythmias in BrS, and body surface mapping and f QRS can be effective predictors of risk in patients with BrS **([1; 2]).** f-QRS appears to be a marker for the substrate for spontaneous VF in BS and predicts patients at high risk of syncope **([3]).**

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Heart rate turbulence (HRT) as predictor of SCD in TOF patients after repair

TOF patients face an increased risk of SCD late after repair. HRT indices are well-known predictors of SCD. Davos et al ([1]) aimed to estimate whether HRT is impaired in repaired T4F patients compared to healthy controls and relate those HRT parameters to already recognized prognostic markers. Continuous ECG recordings were performed in 19 patients late after ToF repair and 20 age-matched healthy controls. Turbulence slope (TS) and onset (TO), frequency and time domain heart rate variability (HRV) parameters and QRS duration were estimated. Volumes of the RV and LV and EF were assessed by cardiovascular magnetic resonance imaging. Cardiopulmonary exercise testing was used to estimate peak oxygen consumption (VO(2)) and VE/VCO(2) slope.

TS and TO were found to be significantly different between TOF patients and controls.

TO correlated with LVEF, LVSVi, RVEF, peak VO(2), VE/VCO(2) slope and with HRV frequency domain indices.

The authors conclude that HRT indices are impaired in T4F patients late after surgical repair compared to healthy controls and relate to coexisting haemodynamic, ventilatory and autonomic impairment. A clinical prognostic role of HRT may be speculated, which warrants further investigation.

1. **Davos CH, Moutafi AC, Alexandridi A, Petropoulou E, Varela E, Chamakou AC, Francis DP, Kilner PJ, Piepoli MF, Gatzoulis MA. Heart rate turbulence in adults with repaired tetralogy of Fallot. Int J Cardiol. 2009 Jul 10;135:308-314.**