



ROMANO - WARD SYNDROME ASSOCIATED WITH TU
ELECTRICAL ALTERNANS: REPORT OF A CASE

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SUMMARY:

A case of the electrical alternans of the TU wave and periodic negative U wave associated with clinical symptoms, electrocardiographic and postmortem findings of Romano-Ward Syndrome has been presented. No electrolyte disturbance was found to be responsible for this exceptionally rare situation. Changes in A-V conduction and left bundle branch block could be attributed to the diffuse coronary sclerosis and subsequent ischemia in the myocardial conduction tissues.

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The electrical alternans of the U wave or TU complex of the electrocardiogram is an exceedingly rare situation without any clearly known mechanism for its appearance. A case of this phenomenon in association with Romano-Ward Syndrome has been presented wherein an abnormality in A-V conduction and left bundle branch block could be encountered.

CASE REPORT

A sixty four-year old woman was referred to the hospital because of frequent syncopal attacks and vague chest pain. She remembered these attacks from the age of 15 for which she had gone under neuropsychiatric investigation (including EEG) and received some medication without any benefit. The patient had not used any cardio-active drug. In her family history, an important point was the sudden death of her two sisters in the youth. (They were only three sibs). Physical examination of the heart and other systems revealed no clinical finding. Blood pressure was 140/90 and temperature 37°C. Chest X ray showed mild basilar pulmonary congestion and moderate cardiomegaly, predominantly the left ventricle.

Laboratory findings were within normal limits:

RBC = 4×10^{12} /lit., WBC = 8000, with normal differential count, ESR = 8^{mm} first hour, 12^{mm} second hour, BUN = 38, FBS = 110, SGOT = 12, and SGPT = 8. CPK, LDH, serum electrolytes (Na^+ , K^+ , Ca^{++} , Mg^{++} , Cl^-), phosphate and urinalysis were normal. In electrocardiogram (as shown below), Q-T prolongation associated with electrical alternans of the T-U wave and periodic negative U waves were noted in the most leads. The P-R interval of 0.24 seconds in most

leads suggesting first degree A-V block and the QRS duration of 0.11 seconds with left bundle branch block pattern ($VAT_{v5,v6} = 0.06$ seconds) were noted in the repeated traces of EKG recorded at different dates. Simultaneous phonocardiogram (not shown here) showed the TU complex appearing later than 0.04 seconds after the beginning of S_2 .

Intracardiac pressures measured by brachial catheterization were as follows:

Right vent. systolic pressure = 29^{mmHg}

Right vent. diastolic pressure = 7^{mmHg}

Pulmonary systolic pressure = 28^{mmHg}

Pulmonary diastolic pressure = 14^{mmHg}

Pulmonary wedge pressure = 15^{mmHg}

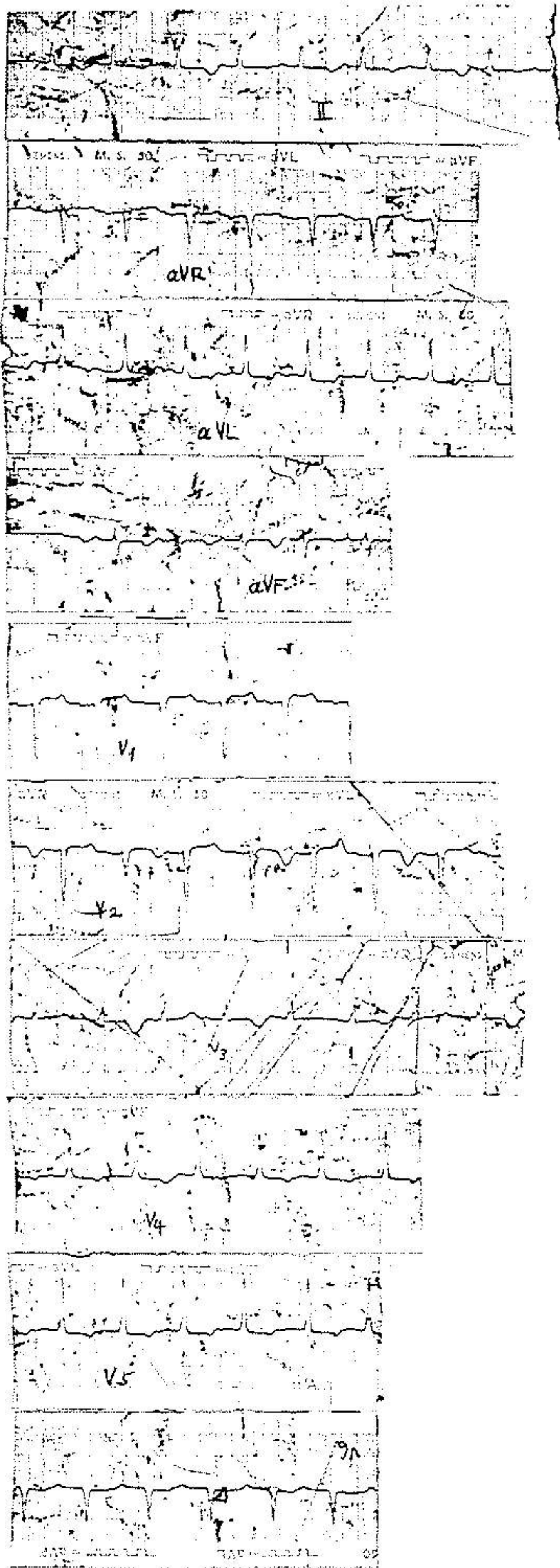
Left vent. end diastolic pressure = 17^{mmHg}

Coronary angiography was not performed.

Intravenous perfusion of CLK, CL_2Mg , and Thromcardin (a mixture of CLK and CL_2Mg) produced no change in E.K.G. The patient did not receive any other drug (including digitalis, antiarrhythmics, etc.).

During two months follow-up of the patient no change in physical and electrocardiographic findings was noted except for an attack of ventricular fibrillation from which she saved by appropriate cardioversion.

The patient died at the age of 64. Postmortem examination revealed diffuse coronary sclerosis both in the main and the small branches. No pathologic finding was noted in the other organs.



P-II (ε₁) =
 P-Form
 QRS - Complex in
 V₁
 V₂
 V₄
 V₆
 Sokolow-Index =
 V₆ = V₁ = mSec.
 ST
 T

Rhythm:
 Heart rate
 P m Sec.
 PQ " "
 QRS " "
 QT " "
 QTc " "
 QRS-Axis (frontal)
 T -Axis (frontal)

DISCUSSION:

Lepeschkin in 1957 described the properties and genesis of the U wave in the electrocardiogram on the basis of the phase 3 repolarisation of Purkinje fibers⁽⁵⁾. Later negative afterpotentials of ventricular myocardium was considered to be the cause of the appearance of the U wave.

Jervell and Lange-Nielsen (1957) reported four members of a family with congenital nerve deafness, prolongation of the Q-T interval, syncopal attacks and sudden death following ventricular arrhythmia.⁽⁴⁾ Romano in 1963⁽¹⁰⁾ and Ward in 1964⁽¹²⁾ described similar patients but with no hearing disturbance and autosomal dominant pattern of transmission. Littmann in 1963 regarded the appearance of the U wave in EKG as an early sign of coronary artery disease and subsequent left ventricular failure.⁽⁸⁾

A few cases of negative U wave have been observed as the consequence of left ventricular hypertrophy and failure, left bundle branch block, complete heart block, coronary artery disease and very rarely in digitalis intoxication, congenital heart disease and cor pulmonale. Q-T or Q-U prolongation produces asynchronous repolarisation of the myocardial fibers.

This in turn results in the appearance of a unidirectional block, producing premature ventricular contraction and finally ventricular tachycardia and fibrillation.

In our case, the familial occurrence of sudden death, syncopal attacks, prolonged Q-T interval and death following ventricular fibrillation in the absence of any electrolyte disturbance and drug overdose confirm the diagnosis of Romano-ward Syndrome. The electrical alte-

rnans of the T-U complex and periodic negative U wave could be seen in the most leads.

According to the previous articles and review of the literature, this combination of Romano-Ward Syndrome and TU electrical alternans has not been yet reported except for the case published by Sano and Hiejima in 1976. Thus our case appears to be the second one reported in the literature.

Is the wave considered as T wave in Jervell-Lange-Nielsen and Romano-Ward Syndromes really T or U wave? Shall the prolonged Q-T interval in those syndromes be considered as Q-U ?

Now it is clear that the prolonged interval formerly known as Q-T in hypokalemia is indeed Q-U.

Though it appears logical to explain the prolonged interval in those syndromes in the term of Q-U interval, the precise answers to these questions remain to be clarified by further electrophysiologic investigations in the future.

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