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SEVERITY OF FALLOT'S TETRALOGY AND ITS RELATIONSHIP TO ELECTROCARDIOGRAPHIC OBSERVATIONS *

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The clinical and hemodynamic spectrum in the tetralogy of Fallot is wide. Prognosis varies with the severity of the anomaly in a patient. Both exercise tolerance and life expectancy in this disease depend on the extent of the pulmonary blood flow; therefore it is important to assess accurately the severity of the lesion.

Dextroposition of the aorta, and existing ventricular septal defect (generally only 1 to 2.5 cm in diameter) are ordinarily not determining hemodynamic factors in Fallot's tetralogy.[1] The volume of blood reaching the pulmonary and systemic vessels from the right ventricle is dependent on the relative resistance presented by the obstruction of the right ventricular outflow tract and by the systemic circulation. Since systemic vascular resistance neither shows much individual or age-related variation, the determining factor in the severity of the anomaly is usually the degree of pulmonary stenosis. The more severe the stenosis, the lower the blood flow form the right ventricle to the lungs and the greater the amount of right to left shunt. In severe cases of Fallot's tetralogy, it has been observed that the pulmonary blood flow is diminished as much as 20 to 30 per cent of the total systemic venous return.[2] In such cases, the left atrium and the cavity of the left ventricle remain hypoplastic.[3-7] In an

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effort to compensate for the reduced pulmonary blood flow and anoxemia, the collateral circulation develops and increases with age. Yet, even this is inadequate and is usually just enough to maintain life.[1] Further compensatory mechanisms, which are related to the degree of unsaturated oxygen, are reduction in oxygen consumption[8,9] and the development of polycythemia. Cyanosis is a manifestation of the amount of reduced hemoglobin in arterial blood; and its development depends on a diminution in pulmonary blood flow, provided that anemia does not coexist. Pulmonary stenosis is the paramount factor in the reduction in pulmonary blood flow; hence, the onset of cyanosis is early and its intensity is marked in severe cases of Fallot's tetralogy.

Though electrocardiographic observations in the tetralogy of Fallot have been studied by various authors, little emphasis has been given to these observations in relation to the severity of the disease.[2,6] The present study proposes to investigate the relationship between the severity of the disease and the electrocardiographic alterations.

Material and Methods

Eighty-four patients with Fallot's tetralogy were selected for the study. Their records were obtained from the Departments of Medicine and Surgery, Cerrahpaşa Hospital, Medical Faculty, University of Istanbul, and from the Hacettepe Medical Faculty, University of Ankara. The requirements for selection of cases were as follows: 1. established diagnosis, 2. evidence of right ventricular hypertrophy in the electrocardiogram (for the 61 surgery patients obtained shortly before the operation), 3. absence of associated cardiovascular disease or anomalies (such as hypertension, dextrocardia) which might complicate the evaluation of the data.

Surgery was performed in the cases of 61 patients; Blalock-Taussig anastomosis in 45 of them. Right heart catheterization in 33, and angiocardiography in nine patients served to confirm the diagnosis. The cases of three patients were confirmed by necropsy. Fifty-one patients were male and 33 female of ages varying from four months to 25 years with a median age of seven.

The mean QRS axis in the frontal plane was estimated by refering to the hexaxial lead system and standardization was carefully assessed. The amplitudes of the Q, R and S waves in leads V_1 (V_{3R}), V_5 and V_6 were read to the nearest 0.5 mm and the average

value of the three complexes were registered. The following QRS voltage criteria were used for right ventricular hypertrophy: R/S ratio above 1.3 and/or amplitude of R wave over 16 mm in V_1 or V_{3R}.[10]

The grade of cyanosis was estimated according to apparent severity, age of onset, and in 17 patients, arterial oxygen saturation. The grade of polycythemia was estimated by the mean preoperative values of erythrocytes, hemoglobin and hematocrit. Polycythemia and cyanosis were estimated minimal, mild, moderate and severe. The chest roentgenograms were interpreted with regard to pulmonary vascular markings and were also graded as normal (0), or diminished (-1, -2, -3). In cases of patients with available angiocardiograms, the degree of stenosis of the right ventricular outflow tract and the caliber of the branches of the pulmonary artery were weighed heavily; furthermore, the degree of overriding of the aorta, the filling sequence, relative densities and sizes of the pulmonary artery and aorta, and the presence of collaterals were considered.[11]

The patients were classified according to the severity of their condition: 1. the degree of cyanosis and polycythemia, 2. the estimate of radiological pulmonary vascular markings, 3. the angiocardiographic observations, and 4. surgical observations when available (estimated diameter of the pulmonary artery or of its left branch, degree of pulmonary stenosis). Severity grades were assigned to patients according to the definition given in Table 1.

TABLE 1
CLASSIFICATION OF SEVERITY GRADE IN FALLOT'S TETRALOGY

Severity grade	Radiologic vascular markings	Degree of cyanosis and polycythemia	Size of pulmonary artery and main branches
1	not diminished	minimal or mild	within normal limits
1-2	slightly diminished	mild or moderate	close to normal
2	moderately diminished	mild or moderate	hypoplastic
2-3	moderately diminished	moderate or severe	hypoplastic
3	markedly diminished	moderate or severe	markedly hypoplastic

Results

The results pertaining to the relationship of the alterations in the right and left precordial leads to the estimated severity grade of the disease, described in *Material and Methods,* are presented below.

1. Relationship of mean QRS axis to degree of severity

$\hat{A}_{QRS}$ in the frontal plane varied between +96° and +240° with a mean value of +143°. The mean values of $\hat{A}_{QRS}$ in the different groups of severity grade were as follows (from grade 1 to grade 3 in increasing order): +117°, +140.6°, +140.7°, +156° and +153°. There was considerable overlapping of individual values in the various groups. It may be concluded that a mean QRS axis of less than +120 indicates only that the severity is not marked, while a deviation of the QRS axis farther than +145° to the right suggests that the case is more severe.

2. Relation of R wave and R/S ratio in V_1 to the degree of severity

Both the amplitude of R wave and the R/S ratio in lead V_1 showed no significant correlation to the severity grade: $r = -0.07$, $p > 0.05$; and $r = -0.02$, $p > 0.05$, respectively (Figures 1 and 2).

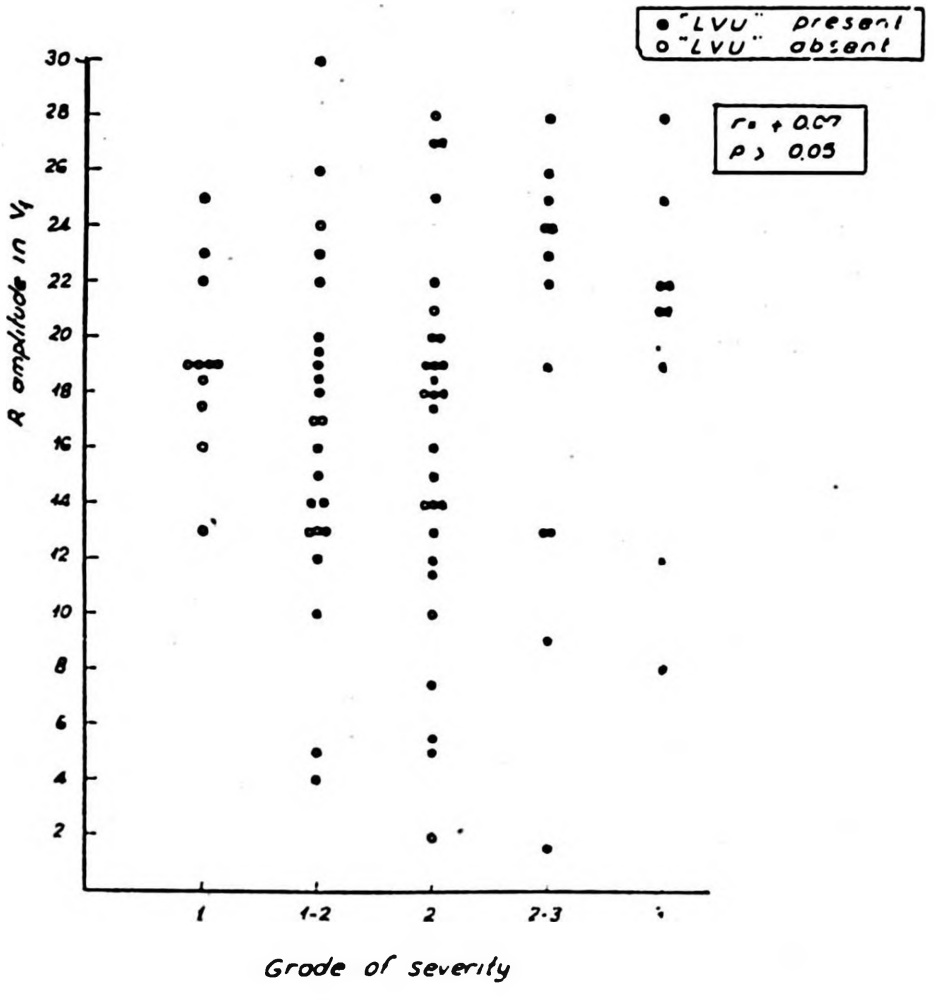

Figure 1. The relationship between the amplitude of the R wave in lead V_1 and the grade of severity in 84 patients with Fallot's tetralogy.

In other words, neither the R wave amplitude, nor the R/S ratio in lead V_1 increased with the increasing severity of the disease.

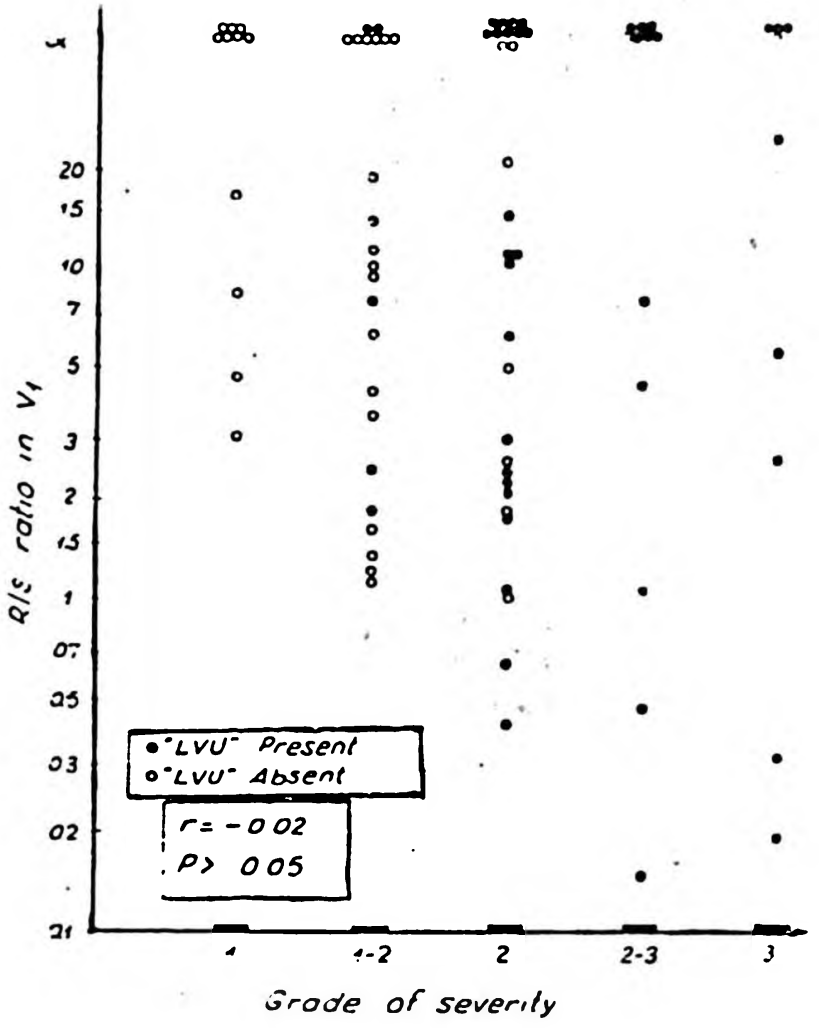

Figure 2. The relationship between the R/S ratio in lead V_1 and the grade of severity in 84 patients with Fallot's tetralogy.

3. Relationship between left precordial leads and grade of severity

a) The relationship between the amplitude of the R wave in lead V_6 and the grade of severity is represented in Figure 3. It is observed that with increasing severity, the R wave in V_6 diminishes ($r = -0.57$, $p < 0.001$).

b) The inverse relationship existing between the R/S ratio in lead V_6 and the grade of severity is presented in Figure 4 ($r = -0.59$, $p < 0.001$).

c) The relationship of the amplitude of the R wave in V_5 to the degree of severity is plotted in Figure 5; the more severe the case, the lower the amplitude of the R wave ($r = -0.55$, $p < 0.001$).

d) The relationship between the R/S ratio in V_5 and the degree of severity is represented in Figure 6, where a statistically higher significant correlation is noted ($r = -0.46$, $p < 0.001$).

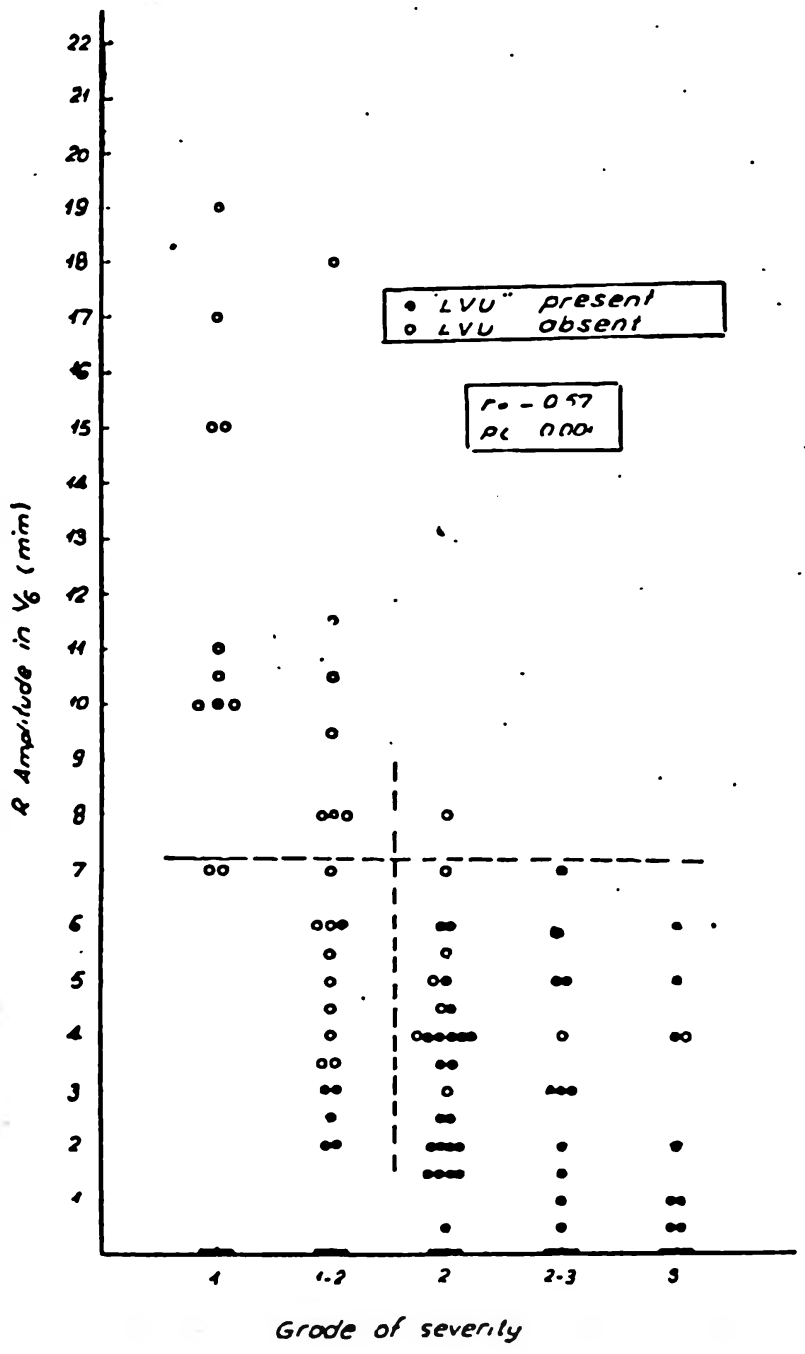

Figure 3. The relationship between the amplitude of the R wave in lead V_6 and the grade of severity in 84 patients with Fallot's tetralogy.

It is thus concluded that with the increasing hemodynamic severity of Fallot's tetralogy, the voltage of the R wave and the R/S ratio in the left-sided precordial leads tend to be lower.

4. Electrocardiographic changes following Blalock - Taussig anastomosis

Due to the consideration that a short period of time after a shunt operation might not allow the development of the left ventricle nor of significant electrocardiographic changes; only those 16

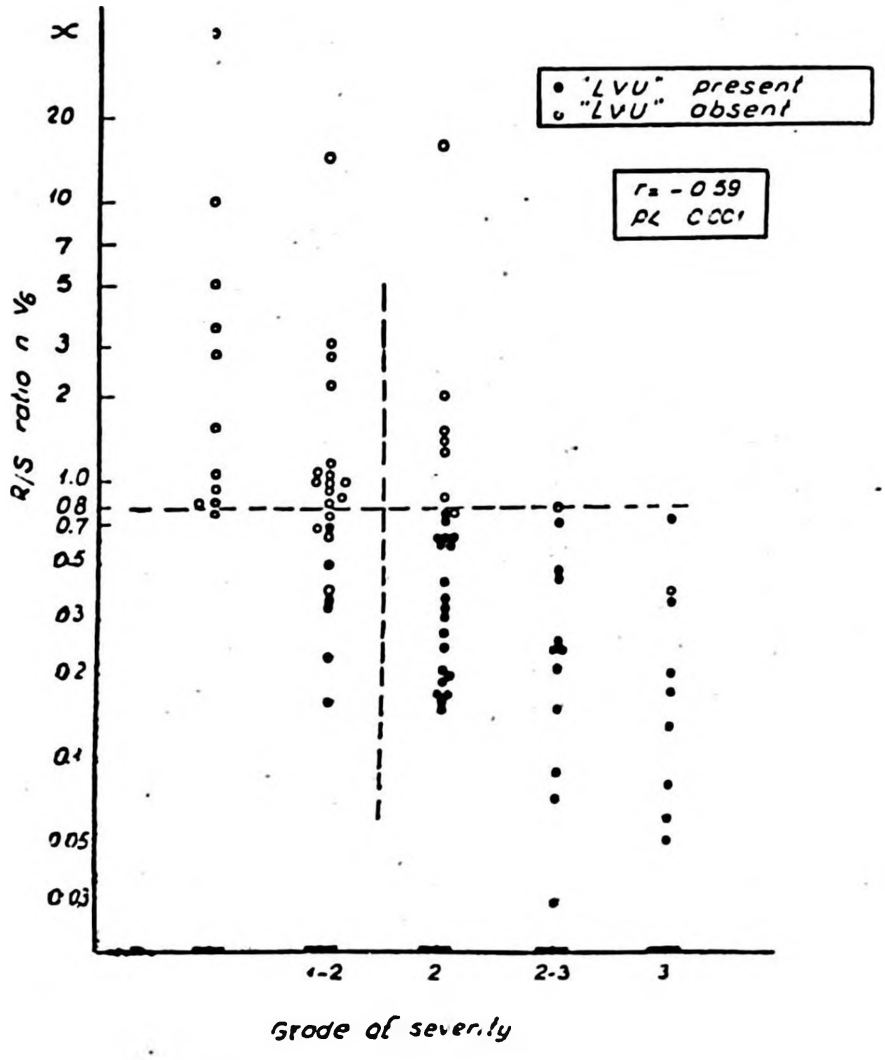

Figure 4. The relationship between the R/S ratio in lead V_6 and the grade of severity in 84 patients with Fallot's tetralogy.

patients who had follow up tracings of more than three months' duration were included in the study. The R/S ratio in lead V_6 increased postoperatively in 11 patients, remained unchanged in one and diminished in four patients; thus the ratio increased from an average of 0.50 to 0.83 (Figure 7a). The amplitude of the R wave in V_6 increased in 14 patients, remained unchanged in one and decreased in

one patient; on the average it increased from 4.28 to 9.44 mm (Figure 7b). Since the postoperative rise in the R/S ratio in the left precordial leads might be due to a reduction in right ventricular hypertrophy, the changes in the voltage of the R wave in V_1 were also studied (Figure 7c). The R wave in V_1 increased in amplitude in the

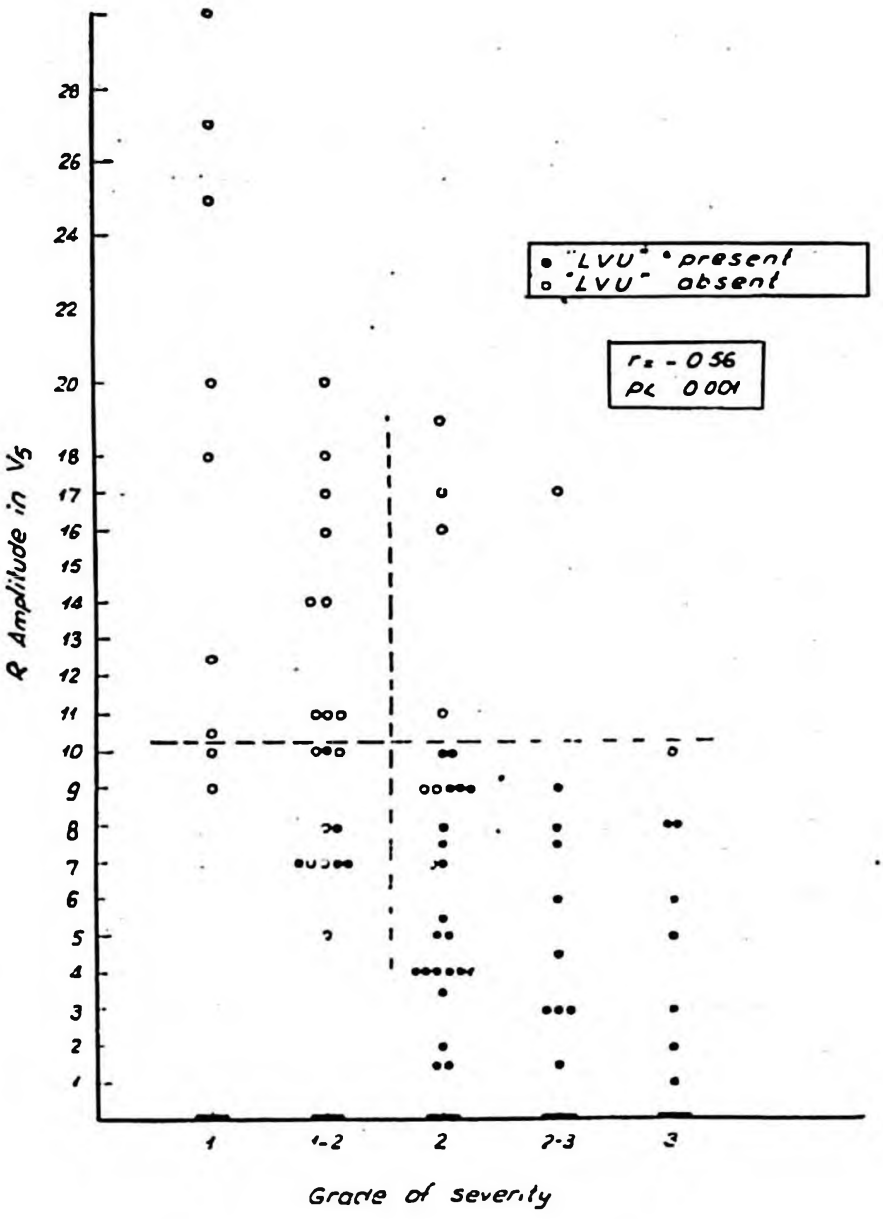

Figure 5. The relationship between the amplitude of the R wave in lead V_5 and the grade of severity in 84 patients with Fallot's tetralogy.

tracings of 12 patients, remained unchanged in two, and decreased in two patients. The mean amplitude of the R wave in lead V_1 did not show a significant rise (15.2 to 18.3 mm), indicating that the postoperative rise in the R/S ratio was not due to a reduction in right ventricular hypertrophy.

5. Q wave in left precordial leads

In 32 of the 84 tracings of patients with Fallot's tetralogy, a Q wave (ranging from less than 0.5 mm to 3 mm in depth) was present in leads V_5 or V_6. A Q wave was recorded in either of these leads in 44 % of patients belonging to the groups with severity grade 1, 1-2 and 2; while it was present only in 13 % of cases with severity grade 2-3 and 3. The less frequent occurrence of Q waves is probably due

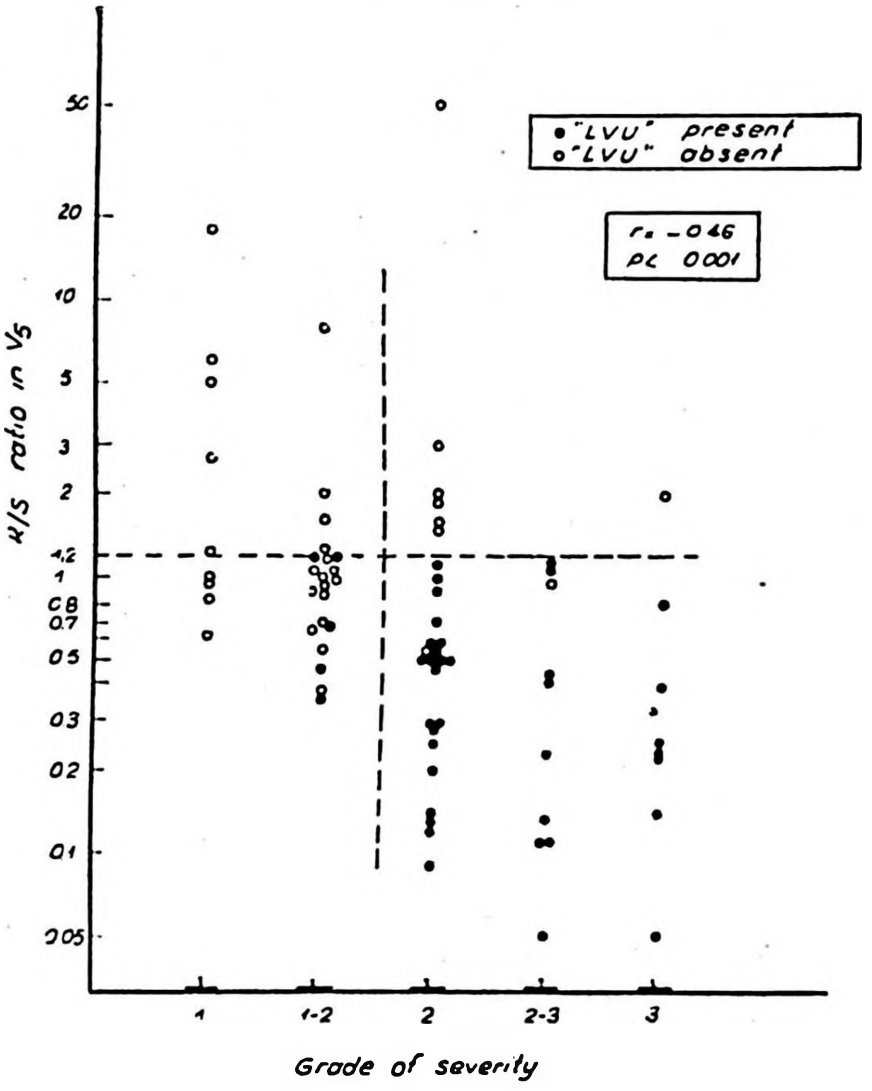

Figure 6. The relationship between the R/S ratio in lead V_5 and the grade of severity in 84 patients with Fallot's tetralogy.

to a more marked clockwise rotation of the heart in severe cases of the tetralogy of Fallot.

6. Electrocardiographic criteria suggesting degree of severity of tetralogy

It seems justifiable to characterize the group of cases with severity grade 1 and 1-2 as «mild» and the group with a higher

degree of severity as «severe». Such a classification, though somewhat arbitrary, permits the establishment of electrocardiographic criteria differentiating the two groups. These criteria were selected from the data presented in Figures 3-6 so that we could get the most sensitive and specific reading. The optimal criteria were as follows:

Lead V_6: R amplitude 7 mm or below and
R/S ratio < 0.8

Lead V_5: R amplitude 10 mm or below and
R/S ratio 1.2 or smaller

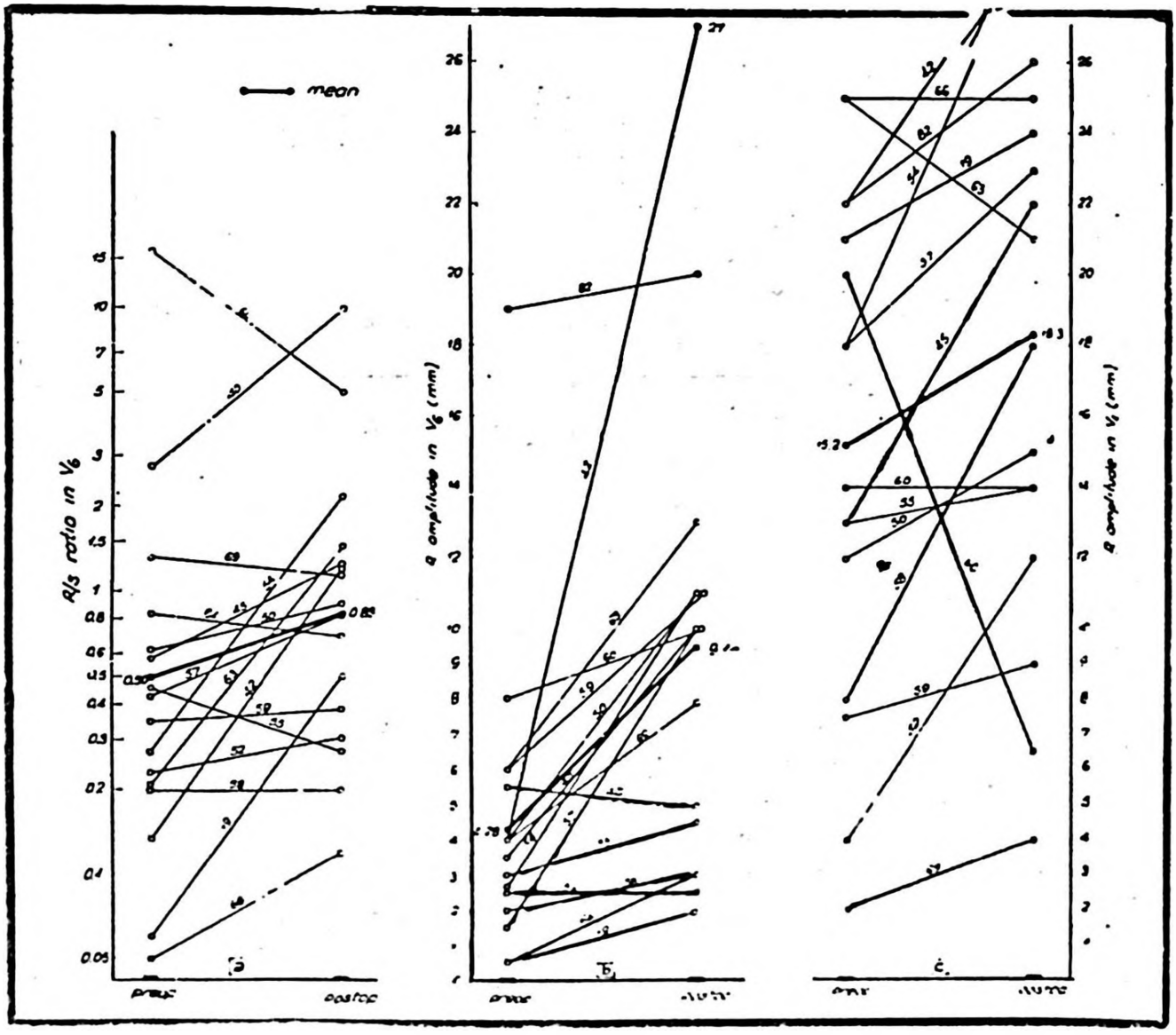

Figure 7 a, b, c. Electrocardiographic changes following Blalock-Taussig anastomosis in 16 patients with tetralogy of Fallot.

Forty-one of the 47 cases meeting these criteria beonged to the group of «severe» cases. In other words, 87 per cent of the patients

with Fallot's tetralogy in which these criteria are met, may be expected to have a severe degree of the disease. On the other hand, among 50 patients comprising the group with «severe» disease, these electrocardiographic criteria were encountered in 41. This indicates a sensitivity rate of 82 per cent.

Discussion

In patients with the tetralogy of Fallot, the main hemodynamic effect is known to be a reduction in pulmonary blood flow, including that portion supplied by collateral circulation which in turn determines the left ventricular inflow. This is the chief factor which determines the degree of severity of the condition which otherwise exhibits a wide divergence with respect to prognosis and ventricular loading.[13] We consider that the severity grade of the disease as defined here is an index of left ventricular inflow. Estimates of roentgenographic and angiocardiographic lung vascularity, of cyanosis and polycythemia, and of the degree of stenosis of the pulmonary outflow tract were considered to reflect the grade of severity of the disease. The study of radiologic lung vascularity in the tetralogy of Fallot, is suitable to estimate approximately the pulmonary blood flow,[6, 14] and, for the purpose of this study, has the advantage of supplying information also on the extent of collateral circulation.[7] Although each of the factors considered in an estimation of the degree of severity has its shortcomings, the fact that a variety of factors has been considered, tends to reduce the sources of error.

The relationship of the amplitude of the R wave and the R/S ratio in leads V_5 and V_6 to the severity grade of the disease was statistically highly significant. Though there was a relatively wide range of values in each grade of severity, the R wave and the R/S ratio over the left precordium tended to diminish with the increasing severity of the disease. On the other hand, no similar relationship was observed between the severity grade and either the R amplitude or the R/S ratio in V_1, signs of right ventricular hypertrophy. These observations agree with the pathologic physiology of the tetralogy of Fallot, since a severe case would have a higher resistance in the right ventricular outflow tract, and a greater amount of blood shunted to the aorta. Consequently, the greater the severity of Fallot's tetralogy, the smaller the diastolic filling of the left ventricle, while the pressure in the right ventricle remains essentially the same. On the basis of these considerations one can conclude that the degree of

severity is reflected by the left rather than by the right precordial leads.

Of special interest was the investigation of electrocardiographic changes following systemic - pulmonary anastomosis, since this type of surgery promotes left ventricular inflow without reducing the pressure load of the right ventricle. We noted a consistent increase in roentgenographic lung vascularity as well as in cardiac size. In 16 patients who were observed for an average of 17 months after Blalock - Taussig anastomosis, the R amplitude and R/S ratio in leads V_5 and V_6 increased significantly, whereas the voltage of the R wave in V_1 did not diminish. These observations are consistent with the better development of the left ventricle postoperatively. Paul et al[15] also observed a slight to moderate increase in cardiac size following Potts' anastomosis in a great majority of their patients which they attributed to the better development of the left ventricle. They reported the frequent appearance postoperatively of electrocardiographic evidence of combined ventricular hypertrophy postoperatively.

Summary

Electrocardiographic observations were investigated in the cases of 84 cyanotic patients with Fallot's tetralogy in which the pulmonary blood flow was reduced to varying extents to find the relationship to the degree of severity of the disease. The voltage of R waves and the R/S ratio in leads V_5 and V_6 showed an inverse relationship to the severity grade ($p < 0.001$). On the other hand, no parallel was noted between the R amplitude and R/S ratio in lead V_1, conventional criteria of right ventricular hypertrophy. Optimal electrocardiographic criteria were presented which aid in differentiating «mild» from «severe» cases of the tetralogy. The specificity and sensitivity rates of these criteria were found to be high enough to justify their practical use.

The observations in left - sided precordial leads demonstrated a tendency to regress following systemic - pulmonary anastomosis, a procedure which, by increasing the pulmonary blood flow, promotes left ventricular filling without reducing the pressure load of the right ventricle.

The electrocardiographic criteria are useful as an aid in assessing the severity grade of Fallot's tetralogy and in determining the prognosis.

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