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Trilogy of Fallot: Analysis of 10 Cases Undergone Open Heart Surgery*

Argun Saylam, M.D. / Coşkun İkizler, M.D.****
Yurdakul Yurdakul, M.D.* / Aydın Aytaç, M.D.******

The combination of pulmonary stenosis, right ventricular hypertrophy and interatrial communication is defined as the "Trilogy of Fallot". This clinical entity was first described by Fallot in 1888.[1, 2, 3] This combination is an uncommon type of congenital cardiac anomaly being present only in 16 instances in the series of 1000 congenital cardiac diseases reported by Maude Abbott (1.6 %), as cited by Selzer et al.[4] Danielson et al.[5] presented 83 cases of trilogy among 221 cases of pulmonary stenosis that were operated on (37.5 % of the cases with pulmonary stenosis). Callahan et al.[6] reported the latter ratio as being 33.3 %.

Material and Methods

10 cases of Trilogy of Fallot have been operated on, using open heart surgery between the years 1962 and 1973 in the Department of Pediatric Thoracic and Cardiovascular Surgery at Hacettepe University (Table I). This number is 13.7 % of 73 cases with atrial septal defects and 21.7 % of 46 cases with pulmonary stenosis subjected to operation during the same period. Six of the children were male and four female. The youngest patient was six and the oldest seventeen years old. Almost all of the patients were admitted to the hospital complaining of dyspnea upon exertion. Cyanosis was present since birth in two cases. Three

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** Resident in the Department of Pediatric Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey.

*** Lecturer in the same department.

**** Professor and Chief of the department.

cases were cyanotic during exercises. Five cases were acyanotic. Squatting was present in two cases, and clubbing in three. In seven patients preoperative diagnosis of Trilogy was made with or without additional cardiac anomalies such as corrected transposition of the great vessels, anomalous pulmonary venous drainage and tricuspid stenosis. In three cases exact diagnosis could not be made preoperatively. These cases were diagnosed as: Tetralogy of Fallot, ventricular septal defect and pulmonary stenosis, and pure pulmonary stenosis (Table I).

Values of cardiac catheterization were parallel to the severity of the lesion. Two cases who were cyanotic since birth displayed significantly increased gradients between the right ventricle and the pulmonary artery.

In one of these cyanotic cases (Hospital number: 65/52065) right ventricular and pulmonary arterial pressures were 228 and 18 mmHg, and in the other one (Hospital number: 65/61115) 186 and 24 mmHg, gradients being 210 and 164 mmHg, respectively. Right atrial oxygen saturations increased in both cyanotic and acyanotic groups, being 70-80 %.

Patients were subjected to open heart surgery using the Kay-Cross disc oxygenator or the Rygg-Kyvsgaard disposable oxygenator with DeBakey roller pumps under normothermic conditions. Oxygenators were primed with 1 or 2 bottles of heparinized blood and 20 ml of Ringer's lactate per kg. of body weight. Pulmonary valvular stenosis was relieved by transpulmonary approach in 9 cases. In one case, subvalvular infundibular bands causing right ventricular outflow tract obstruction were removed by right ventricular approach. In all cases right atrial access was achieved for the closure of interatrial communication.

Results

Pulmonary stenosis was valvular in nine cases and infundibular in one case. Bicuspid valve was present in two cases. Interatrial communication consisted of patent foramen ovale in five cases, and atrial septal defect (ostium secundum) in the rest. Tricuspid stenosis suspected preoperatively was observed in one case and relieved by Gerbode dilatator (other additional pathologies diagnosed preoperatively could not be demonstrated to be present). Pulmonary stenosis was severe in cases showing high degrees of pre-operative gradients across the pulmonary valve (Table I).

TABLE I

CASES OF TRILOGY OF FALLOT UNDERWENT OPEN-HEART SURGERY

Case	Age	Sex	Dyspnea	Squatting	Cyanosis	Clubbing	Preop. Diagnosis	Postop. Diagnosis	Pulmonary Stenosis	Interatrial Communication
Y.G. (65/61115)	17	M	+	+	Since birth	+	Tetralogy of Fallot	Trilogy	Valvular	PFO
H.S. (65/52065)	12	M	+	+	Since birth	+	Trilogy Corrected Transposition	Trilogy	Valvular	PFO
Y.T. (61/6903)	11	M	–	–	–	–	Trilogy + Anomalous Venous draigane	Trilogy	Valvular	ASD
G.P. (75430)	12	F	+	–	On Exercise	–	VSD-PS	Trilogy	Valvular	ASD
H.B. (87614)	13	F	+	–	+	+	Trilogy + TS	Trilogy + TS	Bicuspid valvular	ASD
S.S. (67/62204)	11	F	–	–	–	–	Trilogy	Trilogy	Valvular	ASD
V.S. (205690)	14	M	+	–	On Exercise	–	Trilogy + Anomatous Venous drainage	Trilogy	Valvular	ASD
K.G. (65/39063)	6	M	–	–	–	–	PS+ASD(?)	Trilogy	Bicuspid valvular	PFO
A.O. (59/4328)	15	F	+	–	–	–	Trilogy	Trilogy	Valvular	PFO
Y.G. (60/20671)	14	M	+	–	–	–	PS	Trilogy	Infundibular bands	PFO

Abbreviations:

F : Female
M : Male
VSD : Ventricular Septal Defect
PS : Pulmonary Stenosis
PFO : Patent Foramen Ovale
ASD : Atrial Septal Defect (ostium secundum)
TS : Tricuspid Stenosis

Short and long term follow-up of the patients proved no mortality and all displayed normal physical capacity. Cyanosis disappeared after operation. Post-operative arterial oxygen studies showed an increase to normal levels.

Discussion

"Trilogy" or ""Triad" of Fallot is an uncommon combination among the cyanotic and acyanotic congenital heart diseases. Pulmonary stenosis is usually valvular (patent foramen ovale is the commonest form of interatrial communication followed by atrial septal defect of the secundum type in frequency). Secondary changes in the heart consist of concentric right ventricular hypertrophy, subpulmonary stenosis, tricuspidal fibrosis (as seen in one of our cases) and poststenotic pulmonary dilatation.[7] In rare instances, dextroposition of the aorta,[6] right ventricular endocardial sclerosis (Fig. 1)[7] and single coronary artery[8] are the additionally encountered pathologies. Pulmonary stenosis causes an increase in the right ventricular pressure resulting in concentric hypertrophy. A rise in the diastolic pressure of the right ventricle leads to right to left interatrial shunt and central cyanosis

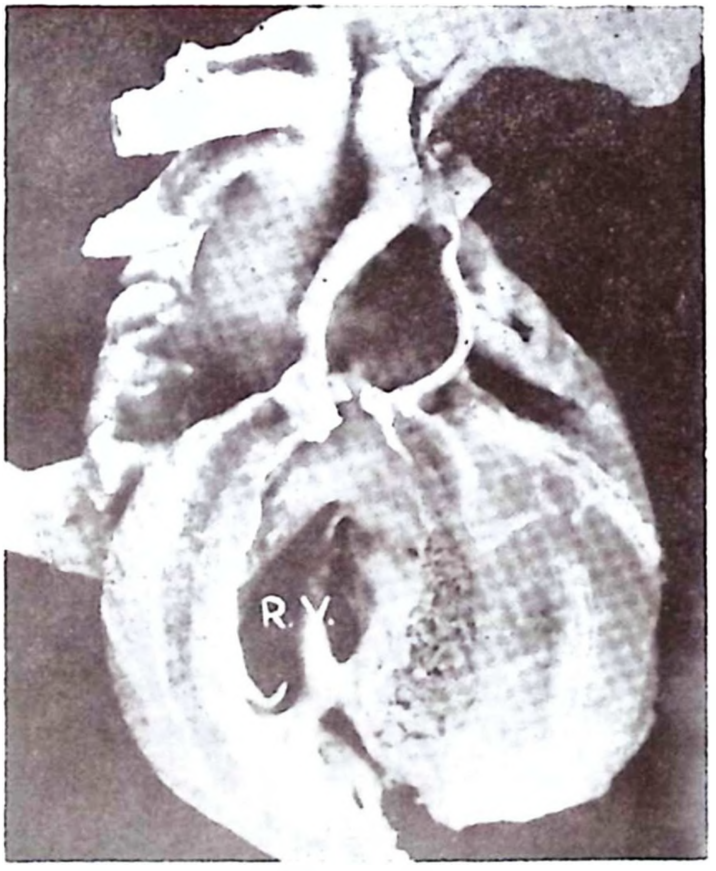

Figure 1

Subendocardial sclerosis in the newborn (Edwards J. E. et al:. Congenital Heart Disease, Volume II. W. B. Saunders, Co., 1965)

(Fig. 2).[7] Magidson et al.[9] stated that in cyanotic cases the gradient across the pulmonary valve is more than 90 mmHg and systemic/pulmonary flow ratio greater than 1/1. In acyanotic cases this gradient is less than 90 mmHg and the systemic/pulmonary flow ratio is smaller than 1/1. Patients are usually cyanotic when pulmonary stenosis is severe.

Dyspnea, cyanosis, clubbing of the fingers, polycythemia, intolerance to exercise, growth retardation, angina pectoris and syncope are the encountered clinical features. Acyanotic patients may be symptomless or dyspnea may be the only predominant clinical feature in such cases.[2, 3, 7] Facies lunata, arachnodactyl and hypertelorism can be seen in rare cases.[3] Auscultation reveals systolic ejection murmur in the pulmonic area which can be inaudable in infants.[10] Cyanosis may persist since birth or take place afterwards (late cyanosis). Late cyanosis occurs when a closed foramen ovale becomes patent due to increased right atrial pressure; thus an acyanotic infant may become cyanotic later in his life. Wood[3] reported that 12% of the presented cases show late cyanosis.

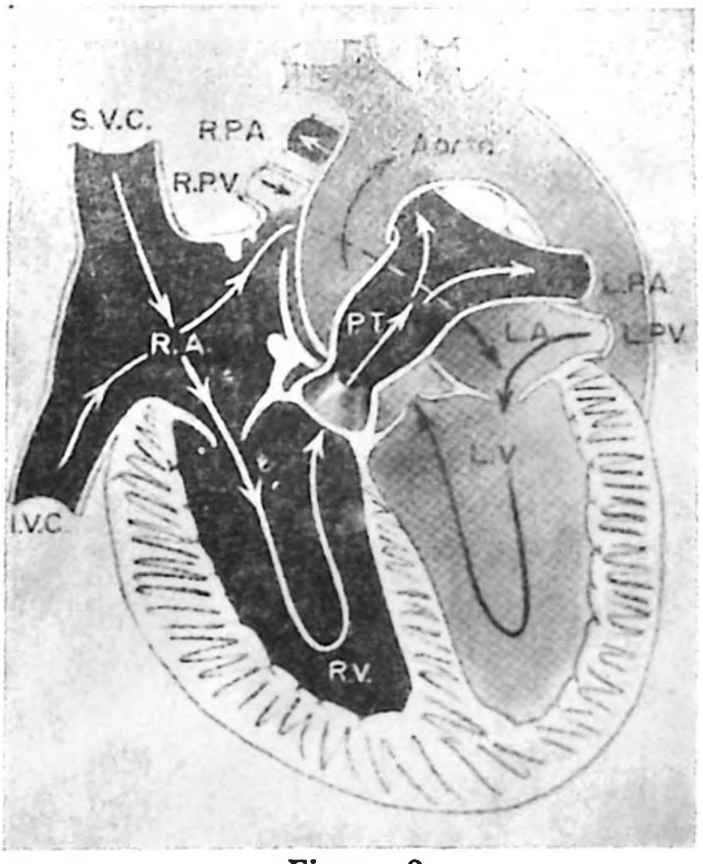

Figure 2

Schema showing hemodynamic alterations in cases of trilogy (Edwards, J. E. et al.: Congenital Heart Disease, Volume II, W. B. Saunders Co., 1965)

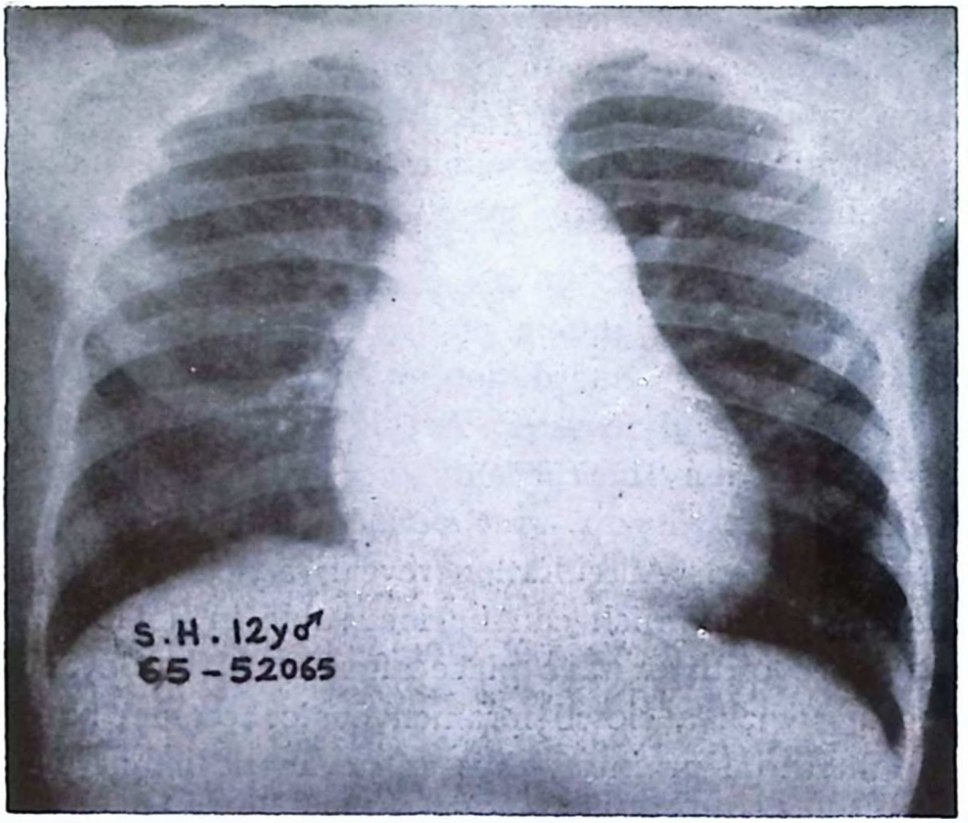

Figure 3
Postero-anterior direct chest x-ray of a patient displaying cyanosis on birth.

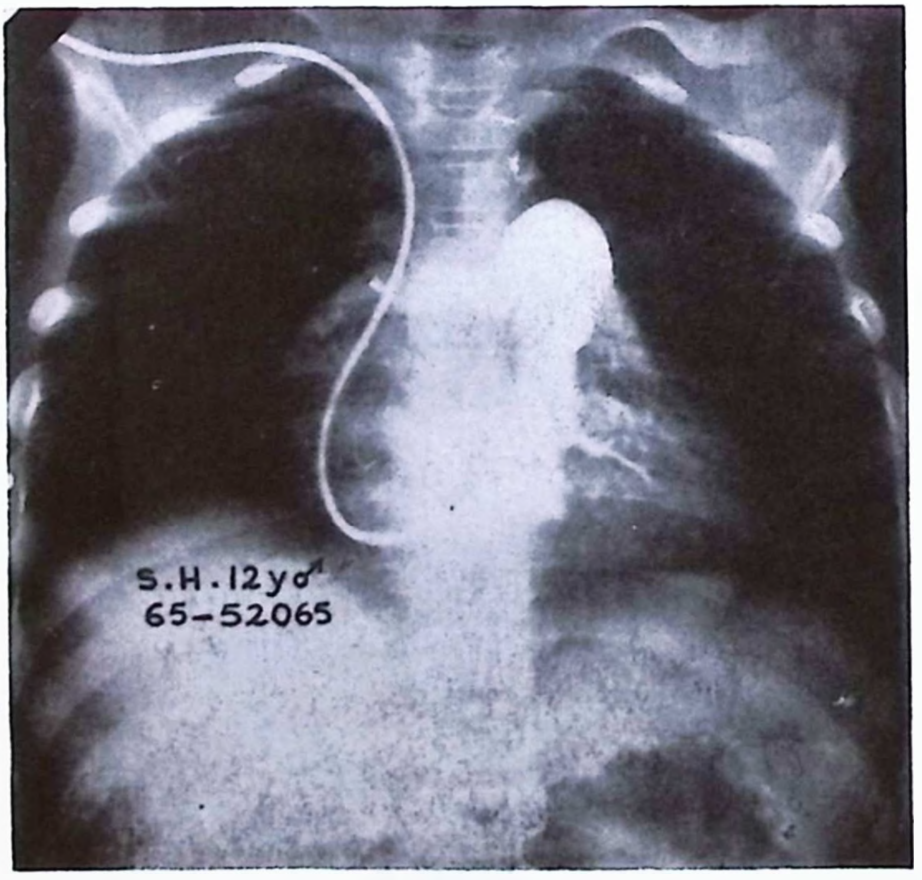

Figure 4
Postero-anterior right heart angiography of the same patient demonstrating a post-stenotic dilatation in the pulmonary artery.

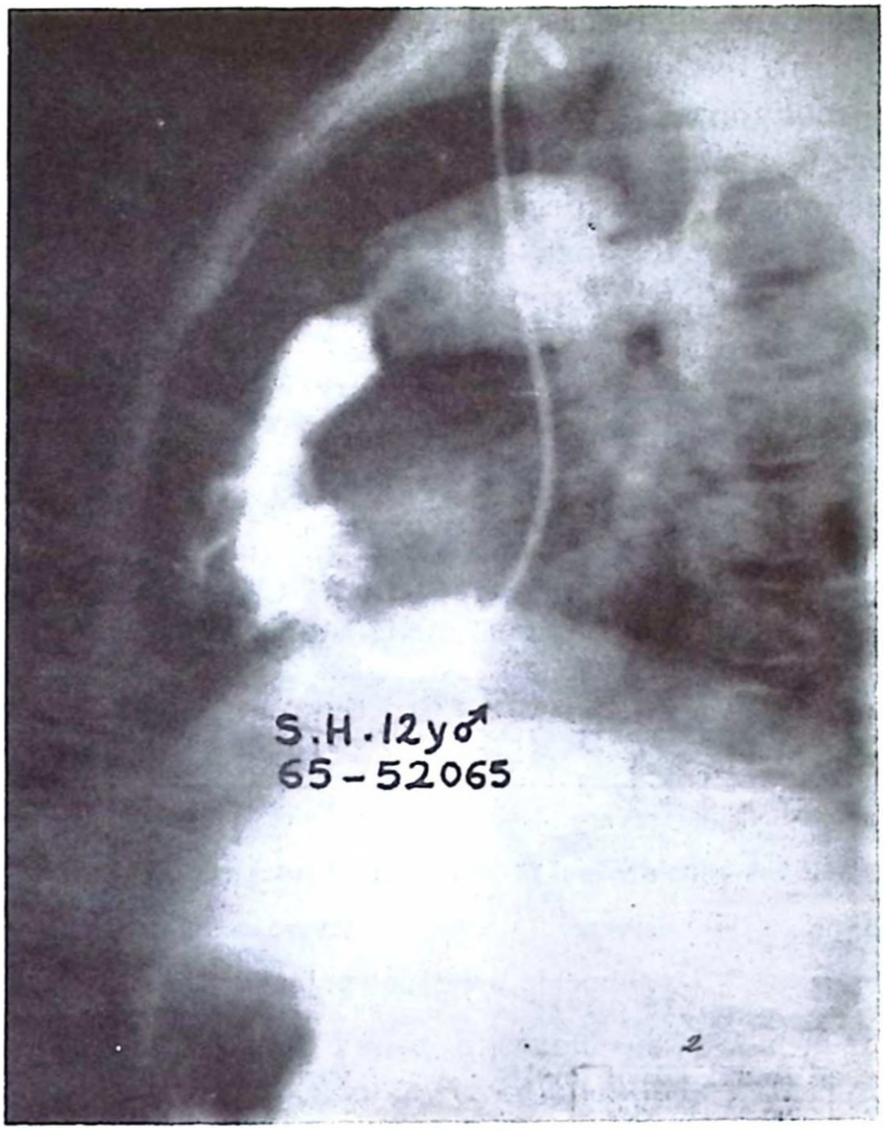

Figure 5

Lateral view of the angiography of the same patient showing the poststenotic dilatation and valvular pulmonary stenosis.

Venous tracings show a giant "A" wave in cyanotic cases. Right axis deviation, right ventricular hypertrophy and right atrial dilatation are recorded in ECG.[3, 7] Direct chest X-rays demonstrate decreased pulmonary vascularity, right ventricular hypertrophy and right atrial dilatation. Angiocardiography may show evidence of poststenotic dilatation in the pulmonary artery (Figs. 3, 4 and 5). Catheterization and angiocardiographic studies are the most helpful diagnostic aids. Differential diagnostic criteria between cyanotic and acyanotic cases are tabulated in Table II. Tetralogy of Fallot, Ebstein's malformation and Eisenmenger's syndrome are to be considered in the differential diagnosis of cyanotic cases (Table III).

Patients may experience hypoxia, cardiac failure, bacterial endocarditis and cerebral abscess (in right to left shunts) as complications.[7]

Modern treatment in cases of Trilogy achieved today is the total correction of this malformation with the help to extracorporeal circulation. Transpulmonary access to relieve the stenosis in the pulmonary valve, and right atrial approach to close the interatrial communication are the techniques used. Relief of pulmonary stenosis is usually effective in infants and patent foramen ovale closes spontaneously after the hemodynamic correction.[10] In the past, inflow occlusion techniques had been performed to correct this pathology. Recently, these techniques have been abandoned and open-heart surgery has become the method of choice to correct these malformations.

TABLE II

DIFFERENTIAL DIAGNOSTIC CRITERIA IN CASES OF CYANOTIC TRILOGY, TETRALOGY OF FALLOT AND EISENMENGER'S SYNDROME

	Cyanotic Trilogy	Tetralogy of Fallot	Eisenmerger's Syndrome
Cyanosis	severe	severe	mild
Clubbing	obvious	obvious	not obvious
Polycythemia	severe	severe	mild
Systolic murmur (localization of)	pulmonic area	pulmonic area	varies
Pulmonic second sound	not loud	varies	loud
Pulmonary arterial shadow in X-rays	large (poststenotic)	small	large
Hilar shadow in x-rays	mildly increased	diminished	increased and pulsatile
Pulmonary congestion	absent	absent	increased
Catheterization and angiographic findings:	a. pulmonary stenosis b. right to left shunt (interatrial)	a. pulmonary stenosis b. Right to left shunt (interventricular) c. dextroposition of the aorta	right to left shunt in various lesions (PDA, ASD, VSD)

Abbreviations:

PDA: Patent ductus arteriosus Botallii

VSD: Interventricular septal defect

ASD: Interatrial septal defect

TABLE III

DIFFERENTIAL DIAGNOSTIC CRITERIAE BETWEEN CYANOTIC AND ACYANOTIC TYPES OF TRILOGY*

	Cyanotic form	Acyanotic form
Pulmonary stenosis	moderate or severe	mild
Cyanosis	present	absent
Fatigue	present	absent
Clubbing	present	absent
Cardiac enlargement	obvious	not obvious
Giant "A" wave	present	absent
P wave in ECG	tall	normal
Pulmonary vascularity	diminished	normal
gradient between the right ventricle and pulmonary artery	more than 90 mmHg	less than 90 mmHg
Left atrial pressure	diminished	normal
Systemic/pulmonary flow ratio	more than 1/1	less than 1/1

* From: Magidson, A. et al.: Am. J. Med. 17: 311, 1954 with necessary variations made.

Summary

Ten cases of Trilogy of Fallot operated on between 1962 and 1973 with the help of extracorporeal circulation were presented. Long term follow-up of these patients proved that surgical treatment performed in these cases were completely successful and effective. Diagnosis and treatment of this malformation were briefly discussed.

Addendum

Two more cases underwent corrective operations after the preparation of this manuscript. The first one was a 12 year-old boy (A.G., 357134) and the second one was a 12 year-old girl (N. Y., 66/13501). Both cases were cyanotic, the first one being cyanotic since birth. Clubbing was present in the first case. Valvular pulmonary stenosis and patent foramen ovale were predominant lesions in both cases. These two children completely recovered.

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