

Clinical Research

Surgical Results After Total Transatrial/Transpulmonary Correction of Tetralogy of Fallot

NICOLAOS M. GIANNOPOULOS, ANDREW C. CHATZIS, ALEXANDROS I. TSOUTSINOS, DIMITRIOS BOBOS, KONSTANTINOS KONTRAFOURIS, MICHAEL MYLONAKIS, GEORGE KIRVASSILIS, IOANNA SOFIANIDOU, PRODROMOS ZAVAROPOULOS, GEORGE E. SARRIS

Department of Pediatric and Congenital Heart Surgery, Onassis Cardiac Surgery Center, Athens, Greece

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Address:
 Nikolaos M.
 Giannopoulos

*Onassis Cardiac
 Surgery Center
 356 Sygrou Ave
 17674 Kallithea
 Athens, Greece
 e-mail:
nickgia@hol.gr*

Introduction: Surgical repair of tetralogy of Fallot is associated with low early morbidity and mortality. However, there may be late morbidity and mortality due to right ventricular dysfunction. The transatrial/transpulmonary technique may ameliorate these long-term complications. Here we present the results from our use of this approach.

Methods: A hundred sixty-three consecutive patients (age 6 months to 45 years, median 1.5 years) underwent transatrial/transpulmonary total correction in our department. In 142 patients the main pulmonary artery was augmented by an autologous pericardial patch, in 31 cases the arterioplasty was extended to the pulmonary artery branches, and pulmonary artery valvuloplasty was needed in 129 patients. A monocusp autologous pericardial valve mechanism was inserted in 14 patients.

Results: Patient follow up was 100% complete with a median duration of 3.05 years. There were no deaths. One patient required early reoperation to relieve residual right ventricular outflow tract (RVOT) obstruction. Median ICU and hospital stay were 3 and 11 days, respectively. At hospital discharge RVOT gradient was 13.7 ± 13 (median 10) mmHg, while most patients (94%) had up to moderate pulmonary valve insufficiency (1+ in 63.8%, 2+ in 30.6%), and normal (92.6%) or mildly reduced (6.1%) right ventricular function. In 81% some degree of tricuspid regurgitation was noted. One patient required late reoperation for mitral valve repair. All patients are in NYHA class I or II. The degree of pulmonary valve insufficiency remains stable (69.9% with 0-1+ and 24.5% up to 2+). Likewise, tricuspid valve function remains unchanged (96% of the patients had mild or up to moderate regurgitation). There was no significant RVOT obstruction and in most patients (93.2%) right ventricular function was normal.

Conclusion: These results compare very favorably to those reported in the literature. The medium-term findings auger well for future adverse event rates, but long-term follow up is still necessary to confirm them.

According to Van Praagh et al,¹ tetralogy of Fallot is in reality a single pathology, which is the result of hypoplasia or incomplete development of the infundibulum of the right ventricle and which covers a wide spectrum of types of obstruction of the right ventricular outflow tract (RVOT). Its surgical treatment was achieved for the first time by Blalock

and Taussig² by a palliative aortopulmonary shunt. Total surgical correction of tetralogy of Fallot, in one or two stages, has been performed successfully for the last 45 years,³⁻¹¹ but it must be noted that long-term postoperative follow up has found an increase in both morbidity (including the need for reoperation) and mortality.¹²⁻¹⁷ Transatrial/transpulmonary correction of

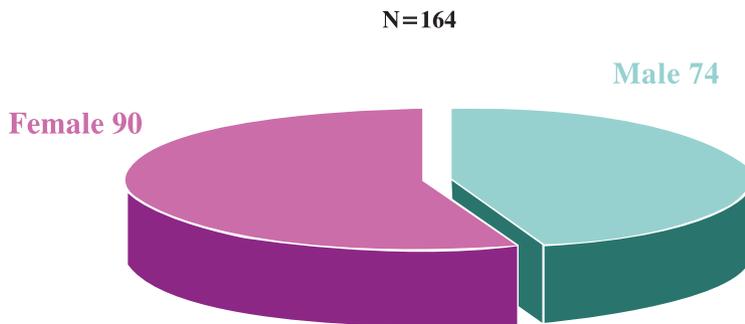


Figure 1. Distribution of patients by sex.

the tetralogy of Fallot was first performed in 1963 by Hudspeth et al⁸ and represented an important development in the surgical treatment of the condition. The method was brought back into the spotlight by Edmunds et al⁶ and gained ever increasing acceptance and application,^{8,10-12,18} with very good results. The theoretical advantages of the method stem from the fact that it provides the best possible preservation of right ventricular structure, geometry and function,^{8,10,19-21} and avoids the undesirable effects^{22,23} associated with long-term right ventricular dysfunction resulting from the classical transventricular technique. On the basis of the above data, the transatrial/transpulmonary approach for total correction of tetralogy of Fallot has been used in our department since its inception. Here we present the immediate and mid-term results.

Methods

Between September 1997 and December 2004 164 consecutive patients (74 male, 90 female, Figure 1), aged 6 months to 45 years (median 1.5 years, mean 3.6 ± 3.4 years) were referred to our department for

surgical treatment, with a diagnosis of tetralogy of Fallot based on echocardiographic investigation with or without cardiac catheterization. The patients' median body surface area was 0.57 m^2 (mean $0.66 \pm 0.2 \text{ m}^2$). The distributions of age and body surface area are shown in detail in figures 2 and 3. Patients with pulmonary artery atresia, absent pulmonary valve or complete atrioventricular canal were not included in this series.

In 163 of these patients total correction was successfully performed using the transatrial/transpulmonary approach. They included 13 out of 14 cases who had previously undergone a palliative aortopulmonary shunt (9 in other centers and 5 in ours). The classification of these 14 patients and the types of shunt are shown in figure 4. Five patients had anomalous origin and course of a coronary artery: in 4 the left anterior descending branch originated from the right coronary artery, while in the other a large branch of the conus artery had a displaced course running around the anterior surface of the pulmonary annulus, ending up parallel to the left anterior descending branch of the left coronary artery (double anterior descending).

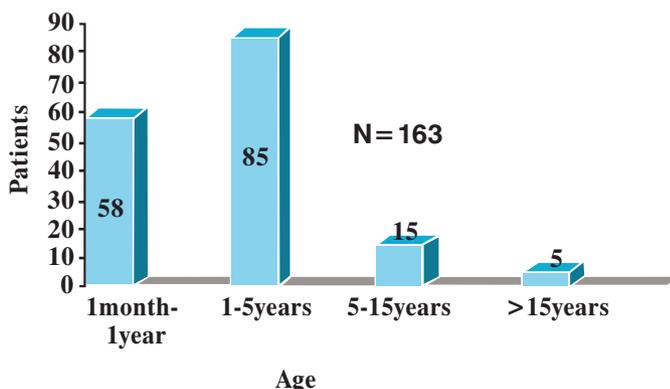


Figure 2. Distribution of patients by age.

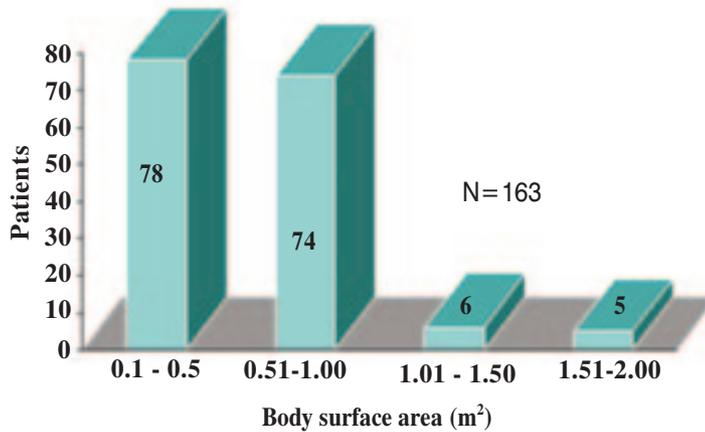
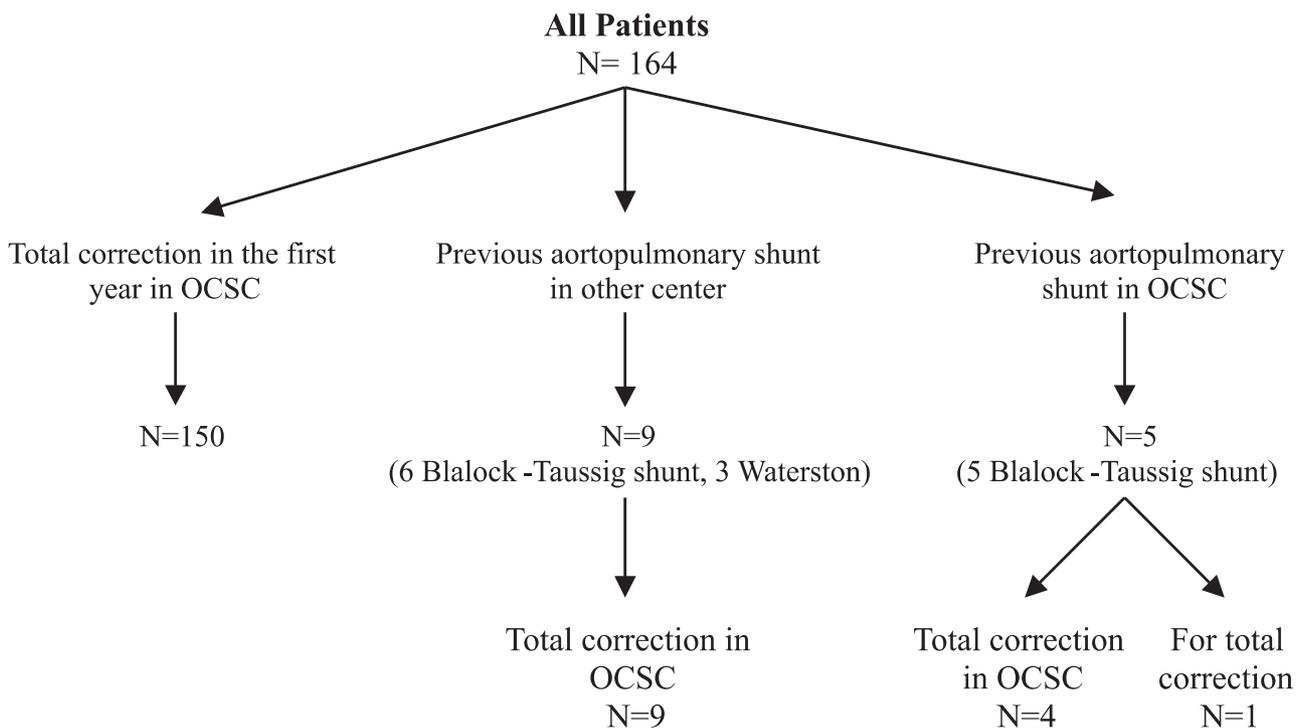


Figure 3. Distribution of patients by body surface area.

The protocol for the transatrial/transpulmonary surgical correction technique that was used in all patients was as follows. Closure of the interventricular septal defect, myectomy of the RVOT and evaluation of tricuspid valve sufficiency (and tricuspid valvuloplasty when necessary) were performed via the right atrium. Completion of right ventricular myectomy,

valvotomy and plasty of the pulmonary valve annulus, arterioplasty of the main pulmonary artery and its branches, when necessary, were carried out via the pulmonary artery. Extracorporeal circulation was applied with venous outflow through catheters in both venae cavae and arterial return through a catheter in the ascending aorta, with maintenance of a moderate



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Figure 4. Analysis of patients based on previous aortopulmonary shunt prior to corrective surgery.

Table 1. Detailed classification of patients according to surgical procedures needed for successful total correction of tetralogy of Fallot.

Procedure	No. of patients
Closure of ventricular septal defect, myectomy of right ventricular outflow tract	163 (100%)
Closure of ventricular septal defect, myectomy of right ventricular outflow tract + plasty of main pulmonary artery	142 (87%)
Closure of ventricular septal defect, myectomy of right ventricular outflow tract + plasty of main pulmonary artery + insertion of small transannular patch (<1 cm)	104 (63.8%)
Closure of ventricular septal defect, myectomy of right ventricular outflow tract + plasty of main pulmonary artery + insertion of large transannular patch (>1 cm)	25 (15.3%)
Closure of ventricular septal defect, myectomy of right ventricular outflow tract + plasty of main pulmonary artery + branch(es)	31 (19%)
Closure of ventricular septal defect, myectomy of right ventricular outflow tract + plasty of main pulmonary artery + insertion of transannular patch + insertion of monocusp valve mechanism	14 (8.5%)

degree of systemic hypothermia (rhinopharyngeal temperature 28 °C). Any preexisting aortopulmonary shunt was then ligated and in the presence of any deformation or stenosis of the pulmonary artery branches it was corrected by autologous pericardial patch. With the heart in diastolic pause, achieved with cold blood cardioplegia and continuous cold pericardial irrigation as described by Shumway, the right atrium was opened with a transverse section. Through this the mural portion of the septum of the infundibulum was divided parallel to the aortic annulus as far as the level of the pulmonary valve annulus. Completion of the myectomy was achieved via transection of the anterior surface of the infundibulum. The ventricular septal defect was then closed with a tailored synthetic Dacron patch, which was sutured with interrupted pledgeted prolene sutures. Tricuspid valve sufficiency was then checked and valvuloplasty performed if it had been deformed as a result of the placement of the Dacron patch. The diameter of the RVOT and pulmonary valve were measured using Hegar dilators. When this was less than the mean normal value according to Rowlatt et al²⁴ the pulmonary artery was opened longitudinally and pulmonary valvotomy was carried out. The aim was to ensure a RVOT diameter 2 mm greater than the mean normal value. If this could not be achieved the pulmonary arteriotomy was extended through the anterior valve crista to the annulus and the right ventricular

wall as far as necessary (usually 0.5-1.5 cm). Autologous pericardium was used to augment the main pulmonary artery, its branches and the right ventricular wall (when necessary). When there were pulmonary artery branches of borderline size (especially in peripheral stenosis) and a significant degree of hypoplasia of the pulmonary annulus, a monocusp autologous pericardial valve was inserted in order to prevent the significant pulmonary valve insufficiency that has been observed in such cases (at least in the immediate postoperative period). The surgical procedures required to achieve total correction are detailed in table 1.

In all patients the right and left ventricular pressures were recorded by direct measurement during surgery and following correction. According to our protocol, when the ratio of right to left ventricular pressure was >75% this would be taken as an indication for further widening of the RVOT: however, such a finding was not seen in any case.

The patients in this series had clinical and echocardiographic assessment in the immediate postoperative period, before discharge from the hospital and on regular follow up visits thereafter. This included recording of the presence and severity of any residual obstruction of the RVOT or pulmonary or tricuspid valve insufficiency, evaluation of right and left ventricular function and checking for any other residual pathology (ventricular septal defect, etc.).

Table 2. Summary of echocardiographic data on hospital discharge.

Right ventricular outflow tract pressure gradient	Mean: 13.7 ± 13 mm Hg	Median: 10mm Hg	
Pulmonary insufficiency	Mean: 0.9 ± 0.9		
Degree	0-1+	2+	3+
No. of patients (%)	104 (63.8%)	50 (30.6%)	9 (5.5%)
Tricuspid regurgitation	Mean: 0.9 ± 0.9		
Degree	0-1+	2+	3+
No. of patients (%)	132 (81%)	31(19%)	0 (0%)
Right ventricular function	Normal	Slightly reduced	Moderately reduced
No. of patients (%)	151 (92.6%)	10 (6.1%)	2 (1.2%)

Results

The mortality in this series of patients was zero, while the median extubation time was 24 hours (6 hours to 27 days, mean 51 ± 35.9 hours), with median ICU stay of 3 days (15 hours to 45 days, mean 5 ± 3 days) and median duration of hospitalization 11 days. The median ratio of right to left ventricular pressure measured during surgery after the correction was 0.57 (mean 0.59 ± 0.41), meaning that in no case was further widening of the RVOT required at that stage. In one patient, however, reoperation was necessary on the fourth postoperative day because of residual RVOT stenosis. That patient, 2 years of age, showed a dynamic type of obstruction due to the particularly long morphology of the right ventricular infundibulum, even though according to the protocol a Hegar dilator 2 mm more than the mean predicted normal size was easily inserted while the heart was in diastolic pause. During reoperation further widening of the RVOT was performed and a monocusp autologous pericardial valve was inserted. The patient's course was uneventful thereafter.

Postoperatively, 18 patients (11%) exhibited transient supraventricular tachycardia, 2 (1.2%) had permanent atrioventricular block and a permanent epicardial pacemaker was implanted before their discharge from the hospital. The remaining patients were discharged in sinus rhythm with a normal QRS complex.

The echocardiographic findings before discharge are shown in table 2. The median pressure gradient between the right ventricle and pulmonary artery during

surgery was 10 (mean 13.7 ± 13) mmHg, with the majority of patients (63.8%) having a mild degree of pulmonary valve insufficiency, while 30.6% had a moderate degree and only 5.5% a severe degree of insufficiency. Tricuspid valve function was well-preserved and in only 19% was there up to moderate tricuspid regurgitation. Right and left ventricular function was good.

Follow up was 100% complete, with a median duration of 3.05 years (2 months to 7 years). During this period only one patient needed reoperation, 5.5 years postoperatively, because of worsening mitral regurgitation (previously undiagnosed cleft in the anterior leaflet, with concomitant pulmonary valve insufficiency and left heart failure. The mitral valve was repaired and a valved right ventricle to pulmonary artery conduit was inserted. The patient's postoperative course was uneventful. All patients are free of symptoms, their electrocardiographic and echocardiographic findings on last follow up examination are summarized in table 3. Specifically, the median pressure gradient in the RVOT was around 14.4 mmHg and no patient developed a severe degree of obstruction during this period, while their right ventricular function remained at a good level. During the same period the rate of preservation of satisfactory pulmonary valve function (69.9% mild and 24.5% moderate insufficiency) and tricuspid valve function (74.8% mild and 21.4% moderate regurgitation) remained high. The comparative echocardiographic findings on discharge and on most recent follow up examination are shown in figures 5 to 8. Finally, the QRS duration remained within normal limits (112.9 ± 22.9 ms).

Table 3. Summary of echocardiographic data on latest follow up.

NYHA Class	I-II	III	IV
No. of patients (%)	163 (100%)	0 (0%)	0 (0%)
QRS duration	Mean: 112.9 ± 22.9 ms		Median: 112 ms
Arrhythmia		No	
Right ventricular outflow tract pressure gradient	Mean: 14.4 ± 12.6 mmHg	Median: 15 mmHg	
Pulmonary insufficiency			
Degree	0-1+	2+	3+
No. of patients (%)	114 (69.9%)	40 (24.5%)	9 (5.5%)
Tricuspid insufficiency			
Degree	0-1+	2+	3+
No. of patients (%)	122 (74.8%)	35 (21.4%)	6 (3.6%)
Right ventricular function	Normal	Slightly reduced	Moderately reduced
No. of patients (%)	152 (93.25%)	10 (6.13%)	1 (0.61%)

Discussion

The surgical correction of tetralogy of Fallot, according to the results of many studies,^{5,9,11,25-27} can be performed with relatively low surgical mortality (0-7%), while there is an increasing trend towards correction at a young age. In spite of this, however, long-term postoperative follow up shows increasing morbidity, since reoperation is needed in 13-20% of patients and in 1-3% more than one repeat procedure is necessary, with a consequent increase in long-term mortality.¹²⁻¹⁶ This undesirable course is attributable to a deterioration in the dilatation and dysfunction of the right ventricle (with tricuspid regurgitation) and at a later phase of the left ventricle,²⁸ re-

sulting in reduced exercise tolerance, arrhythmias and possible sudden death. The mechanisms implicated in this right ventricular distension and dysfunction are a severe degree of pulmonary valve insufficiency, residual or recurring obstruction of the RVOT, aneurysm of the patch used to widen the RVOT, or other residual lesions (ventricular or atrial septal defect, stenosis of pulmonary artery branches).^{21,26-29} The factors that predispose for the appearance of the above mechanisms and are related to the surgical technique include the following:

1. A long ventriculotomy combined with exaggerated myectomy of the RVOT and a large transannular patch, leading to aneurysm of the RVOT;²²

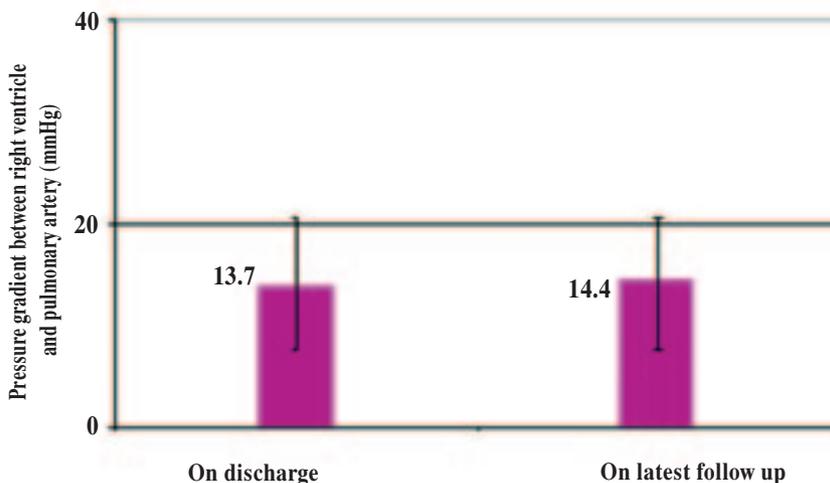


Figure 5. Mean pressure gradient between right ventricle and pulmonary artery.

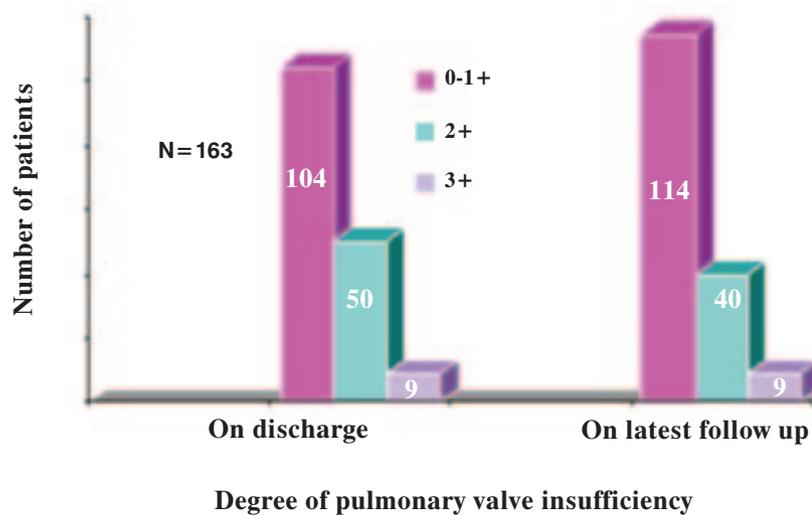


Figure 6. Distribution of patients according to degree of pulmonary insufficiency.

2. A large transannular patch, especially if there is also stenosis of pulmonary artery branches,^{21,22,29} even though its size has not been shown to be an independent factor for increased risk of long-term mortality, is nevertheless associated with an increased probability of reoperation being required, probably because of the anatomy that created the need for a large patch;
3. Residual stenosis of main pulmonary artery and/or branches,^{21,25}
4. The presence of high pulmonary resistances because of previous aortopulmonary shunt;²⁷
5. A high pressure gradient between the right ventricle and pulmonary artery;^{21,22,25,27,29}
6. Use of a transventricular rather than a transatrial/transpulmonary approach to total correction.^{7,11,13-15,22,26,28}

The surgical mortality rate for transventricular correction ranges from 1 to 30% (with higher rates in series from the period 1955-1965), with a reoperation rate 10-20%.^{13-15,22,30} It is notable that in clinical studies using the transatrial/transpulmonary approach the rates of mortality (0-2%) and reoperation (0-5%)^{7,11,30} were significantly smaller than those for the transventricular method, even when the technique was used for all patients, without any selection.¹¹

From a practical point of view, an important aim is the determination of the ideal balance between satisfactory correction of the obstruction of the RVOT and exaggerated pulmonary valve insufficiency. This may be attained by using a patch of "ideal" size, if a patch must be inserted, or two patches using the trans-

ventricular method, or even better, in our opinion, by adoption of the transatrial/transpulmonary approach for total correction of the tetralogy of Fallot.

Our use of transatrial/transpulmonary correction in our department is based on these data and on other data from early series that reported low rates of mortality and morbidity.^{6,7,9,10} Our results indeed confirm that the method is exceptionally safe (zero mortality), but in addition its use also contributes to the preservation of satisfactory right ventricular function, both during the early postoperative period and in the medium term. The echocardiographic data from the patients' discharge examination show that only a small percentage (1.2%) had a moderate reduction in right ventricular contractility, while the overwhelming majority had good or very good right ventricular function.

Furthermore, in this series of patients most (63.8%) had a small degree of pulmonary valve insufficiency, while only 5.5% had a severe degree and in 30.6% the insufficiency was moderate. It should be stressed that the preservation of satisfactory pulmonary valve function was achieved without there being any significant degree of residual obstruction in the RVOT. The median right ventricular pressure gradient after correction, measured during the surgery, was 10 (mean 13.7 ± 13) mmHg and the median ratio of right to left ventricular pressure was 0.57 (mean 0.59 ± 0.41). Another advantage of the transatrial/transpulmonary approach is that the tricuspid regurgitation is checked after the insertion of the patch used to close the ventricular septal defect. The tricuspid valve may then be repaired if it has been distorted, thus avoid-

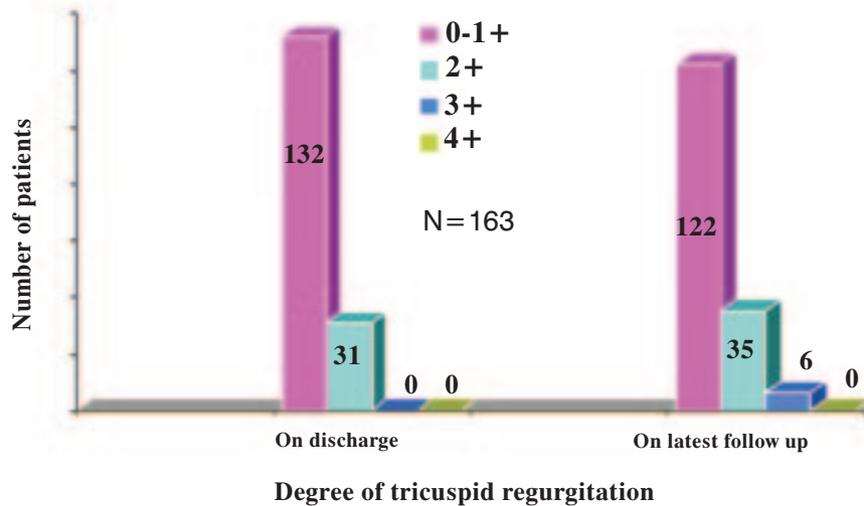


Figure 7. Distribution of patients according to degree of tricuspid regurgitation.

ing a severe degree of regurgitation. Such regurgitation is not at all favorable postoperatively, especially if there is concomitant pulmonary valve insufficiency. In this series no patients showed a severe degree of tricuspid regurgitation during the immediate postoperative period.

Finally, it should be mentioned that the use of the method in cases that have an anomalous course or origin of a coronary vessel reduces the likelihood of the use of a right ventricle-pulmonary artery valved graft. Of the patients included in this series there were five with such a coronary vessel anomaly and no valved conduit was needed in any of them.

It must be stressed that, more important than the immediate postoperative results (which show no signifi-

cant differences between the two methods of surgical correction, although the transatrial/transpulmonary approach is slightly superior to the transventricular) is the patients' course over time, which will become clearer with the long-term collection and comparison of postoperative data. The mid-term clinical and laboratory findings from this series (mean length of follow up 3.05 years, 100% compliance) are as follows:

1. The patients remain free of symptoms, with no need for medication;
2. No significant degree of RVOT obstruction has developed;
3. Pulmonary valve insufficiency remains at moderate levels;
4. Tricuspid valve function remains satisfactory;

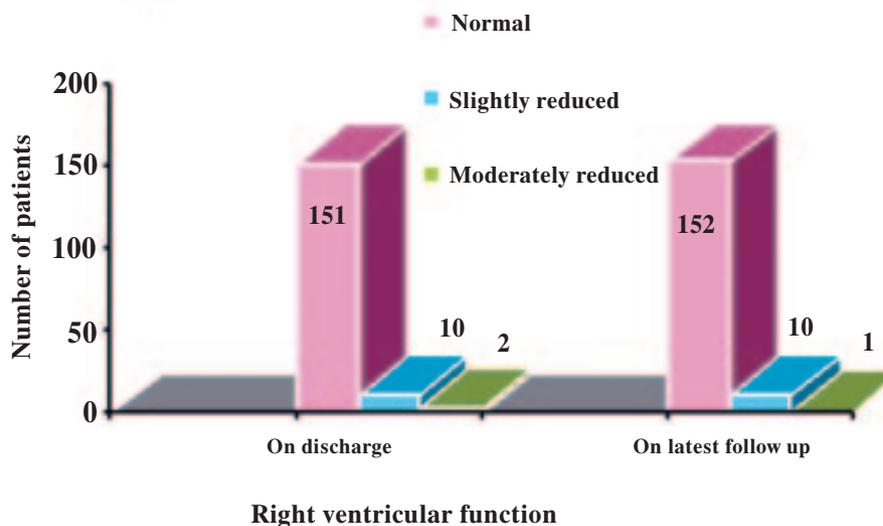


Figure 8. Distribution of patients according to right ventricular function.

5. No arrhythmias have been noted and the QRS duration is within normal limits;
6. Right ventricular function remains very good in spite of the moderate pulmonary valve insufficiency.

These results are encouraging, but the follow up must of course continue for a longer period (up to 10 years) and a more detailed study of right ventricular function will be necessary, for example using magnetic resonance imaging. This is already in progress in our department.

In conclusion, this study shows that transatrial/transpulmonary correction of the tetralogy of Fallot in our department is associated with excellent results immediately following surgery and very satisfactory medium-term clinical and laboratory findings. Of course, further follow up is clearly necessary in order to confirm the initial findings over a longer time, with regard to the preservation of the structure and function of the right ventricle.

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