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EARLY POST-OPERATIVE RESULTS AFTER TOTAL CORRECTION OF TETRALOGY OF FALLOT: THE EXPERIENCE IN SHIRAZ, IRAN

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ABSTRACT

In order to compare early post-operative results in primary versus two-stage repair of tetralogy of Fallot at Shiraz University of Medical Sciences, one-hundred and eleven patients with tetralogy of Fallot with right ventricle to pulmonary artery continuity and no other major associated anomaly were repaired in one center by one surgeon in a one or two-stage protocol. Those patients who were initially palliated with shunt were either referred from other centers for total correction, presented with cyanotic spells, or were shunted due to their severe and diffuse right ventricular outflow tract obstruction or pulmonary artery branch stenosis or hypoplasia. Hospital mortality and ratio of right to left ventricle pressure after correction were compared between the primary and the two-stage groups as early outcome indices. The patients were also divided to those who needed a trans-annular (TAP) or a sub-annular patch and were compared.

In the primary group, 25 (37.3%) of the patients needed TAP, while in the two-stage group 28 (63.6%) needed TAP. Requirement for TAP was increased significantly with two-stage correction (p= 0.006, relative risk= 1.71, 95% CI= 1.16-2.5). Mortality was significantly higher in the primary group (p= 0.03, relative risk= 3.94, 95% CI= 0.93-16.76). In the primary group TAP significantly increased the mortality risk (p= 0.006, relative risk= 5.04, 95% CI= 1.5-16.89). In the two-stage group, there was no significant difference in the mortality rate between the patients with and without TAP. The TAP group had statistically significant less time interval between shunt and total correction.

Our patients generally did better on two-stage repair, because of their older age at operation. The long period of low pulmonary blood flow has induced unbalanced ventricles for them and exaggerated right ventricular outflow tract obstruction due to muscle hypertrophy. In such patients, shunting will prepare the left ventricle for accepting the extra blood volume that will reach it after total correction.

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INTRODUCTION

Total correction of tetralogy of Fallot is a well-established procedure since Lillehei in 1954[13] and Kirklin in 1955[11] performed the first cases. Comparison of the natural history, with an only 10-15% survival after 20 years of age,[1] and total correction with survival and normal life quality of the vast majority of cases after 20-37 years as reported by Norgaard recently,[18] encouraged surgeons today to perform earlier primary repair on patients with more complex forms of the anomaly.

With advances in technology and better post-operative care, primary repair is advocated in all age groups.[3,19,20] This policy needs to be investigated in different centers because of age variability at the time of diagnosis and total correction, technical variability, and difference in post-operative care facility.

The objective of this study was to compare early post-operative results (until discharge from the hospital) in primary versus two-stage repair of tetralogy of Fallot at a Shiraz University of Medical Sciences affiliated hospital between 1991-1999.

MATERIAL AND METHODS

One-hundred and fourteen patients with tetralogy of Fallot with right ventricle to pulmonary artery continuity who were repaired in one center by one surgeon between 1991-1999 were studied. Three of these patients were excluded due to associated anomalies (double chamber right ventricle in two and aortic insufficiency in one).

Of the patients, 67 (60.4%) were primarily corrected and 44 (39.6%) via a two-stage plan. Those patients who were initially palliated with a shunt were either referred from other centers for total correction, presented with cyanotic spell, or were shunted due to to their severe and diffuse right ventricular outflow tract (RVOT) obstruction or pulmonary artery branch stenosis or hypoplasia.

All patients were corrected through median sternotomy with cardiopulmonary bypass, bicaval canulation and moderate hypothemia. The aorta was cross-clamped and cold crystalloid cardioplegic solution given. The VSD was closed mainly through ventriculotomy and occasionally through the right atrium. Release of right ventricular outflow tract obstruction (RVOTO) was performed with longitudinal ventriculotomy. The annulus was sized with a Hegar dilator. If the diameter was more than two standard deviations below the mean normal values established by Rowlatt et al., a trans-annular patch was inserted.[5] The patch was made from untreated autologous pericardium. The proximal extension was stopped or paralleled when it reached a major branch from the right coronary artery to RVOT.

Hospital mortality and ratio of right to left ventricle pressure (RV/LV ratio) after correction were compared between the primary and the two-stage groups as early outcome indices. The patients were also divided to those who needed a trans-annular patch (TAP) or sub-annular patch (RVOT-P) and were compared for hospital mortality and RV/LV ratio.

Two-tailed t-test, single factor ANOVA, chi-square, and Fisher's exact test were used to statistically examine the results, each when appropriate.

RESULTS

Of the 111 patients included in the study 69 (62.2%) were male and 42 (37.8%) were female (M/F ratio= 1.64). In the primary group the number of males was 42 (62.7%) and females was 25 (37.3%). In the two-stage group, it was 27 (61.4%) and 7 (38.6%) respectively (Table I).

The mean±standard deviation (SD) for age in the total patients was 7.65±5.29 years ranging from 4 months to 29 years. This was nearly the same in all groups with a slightly

Table I. Sex distribution and mean and standard deviation for age, weight, and RV/LV ratio in the patients.

	Male	Female	Mean age (year)	SD*	Mean Wt (Kg)	SD	Mean RV/LV ratio	SD
Total	69 (62.2%)	42 (37.8%)	7.65	5.3	19.8	11.7	0.5	0.13
Primary:	42 (62.7%)	25 (37.3%)	7.26	5.2	18.2	11.4	0.5	0.13
Primary TAP			7.24	5.4	19.7	14.7	0.55	0.13
Primary RVOT-P			7.27	5.1	17.1	8.4	0.48	0.13
Two-stage:	27 (61.4%)	17 (38.6%)	8.24	5.4	22.2	12.0	0.5	0.13
Two-stage TAP			7.34	5.5	20.1	9.9	0.5	0.15
Two-stage RVOT-P			9.8	5.2	26+	14.6	0.5	0.14

*Standard deviation
+Significantly higher than the primary RVOT-P group (p= 0.015 using two tailed t-test).

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Table II. Requirement of TAP in the total, primary and two-stage group.

	RVOT-P	TAP
Primary	42 (62.7%)	25 (37.3%)
Two-stage	16 (36.4%)	28 (63.6%)*
Total	58 (52.3%)	53 (47.7%)

*Significantly higher than the primary group (p= 0.006 using chi-square test, relative risk= 1.71, 95% CI= 1.16-2.5).

higher age in the two-stage RVOT-P group with a mean±SD of 9.8±5.24 (Table I). However, age had no significant difference between either of the groups using two-tailed t-test.

The mean±SD for weight in the total patients was 19.8±11.7 kg, ranging from 6 to 65 kg. Except for the two-stage RVOT-P group with a mean±SD of 26±14.6, that was significantly higher than the primary RVOT-P group (p= 0.015 using two tailed t-test), this was nearly the same in all groups (Table I).

Mean pre-operative hematocrit in the primary repair group was 48.7% and in the two-stage group was 51.2% with no significant difference. No coronary artery abnormalities were seen during operation.

RVOT-P was placed in 58 (52.3%) of the patients and TAP in 53 (47.7%). In the primary group, 25 (37.3%) of the patients needed TAP, while in the two-stage group 28 (63.6%) needed TAP (Table II). Requirement for TAP was increased significantly with two-stage correction (p= 0.006 using Chi-square test, relative risk= 1.71, 95% CI= 1.16 - 2.5).

Overall hospital mortality was 12.6% (14 out of 111). There were 12 (17.9%) hospital deaths in the primary and 2 (4.5%) in the two-stage group (Table III). Mortality was significantly higher in the primary group (p= 0.03 using Chi-square test, relative risk= 3.94, 95% CI= 0.93-16.76). Overall, 10 patients (18.8%) with TAP and 4 (6.9%) of those with RVOT-P died. However the differences were marginally insignificant (p>0.05 [p= 0.058] using chi-square test). Considering the primary group alone, 9 patients (36%) with TAP and 3(7.1%) of those with RVOT-P died (Table III). In this group TAP significantly increased the mortality risk (p= 0.006 using two tailed Fisher's exact test, relative risk= 5.04, 95% CI= 1.5.- 16.89). Considering the two-stage group alone, 1 patient (3.5%) with TAP and 1 (6.25%) with RVOT-P died (Table III). There was no significant difference in the mortality rate between the patients with and without TAP in this group.

The mean±SD for RV/LV ratio was 0.50±0.13. It was 0.57±0.14 in patients who didn't survive and 0.50±0.13 in surviving patients. The difference was not statistically significant (p= 0.085 using two-tailed t-test). Arranging the patients in groups with increasing RV/LV ratio (less than 0.5, 0.5-0.75 and equal or greater than 0.75), the mortality

Table III. Mortality rate in different groups.

	Survived	Expired
Total	97 (87.4%)	14 (12.6%)
Primary	55 (82.1%)	12 (17.9%)*
Primary TAP	16 (64%)	9 (36%)+
Primary RVOT-P	39 (92.8%)	3 (7.1%)
Two stage	42 (95.5%)	2 (4.5%)
Two stage TAP	27 (96.5%)	1 (3.5%)
Two stage RVOT-P	15 (93.75%)	1 (6.25%)

*Significantly higher than the two-stage group (p= 0.03 using chi-square test, relative risk= 3.94, 95% CI= 0.93-16.76).
+Significantly higher than the primary RVOT-P group (p= 0.006 using two tailed Fisher's exact test, relative risk= 5.04, 95% CI= 1.5-16.89).

rate increased with rising RV/LV ratios, however not significantly (Fig. 1). Mean and SD of RV/LV ratio for different groups according to the type of correction are summarized in Table I. The RV/LV ratio had no significant difference between either of the groups using two-tailed t-test.

In the two-stage group, mean time interval between shunt and total correction was 43 months (range 1-240) with SD= 40.35. There was no correlation between time interval and RV/LV ratio (correlation coefficient= -0.05). Mean time interval between shunt and total correction was 29.1 months with SD= 16.0 in the TAP group, and 70.81 months with SD= 58.28 in the RVOT-P group. The TAP group had a statistically significant less time interval between shunt and total correction than the RVOT-P group (p= 0.003 using two tailed t-test). Excluding the patient with an abnormally high interval of 240 months from the RVOT-P group, the mean time interval between shunt and total correction was 59.5 months with SD= 16.6, still significantly higher than the TAP group (p= 0.003 using two tailed t-test).

The mortality rate slightly increased along these nine years of 1991-1999 (Table IV), but the changes were not statistically significant (p= 0.9 using chi-square test). Mean age of patients operated in different years was not significantly different (p= 0.2 using single factor ANOVA test), but mean weight slightly increased with statistical significance (p= 0.02 using single factor ANOVA test). The RV/LV ratio was statistically the same in different years of operation (Table IV). The percentage of patients that needed TAP increased along the years of study (p= 0.03 using chi-square test).

DISCUSSION

It is generally accepted that a high postoperative RV/LV ratio is associated with increased hospital mortality.[2,6-8] Murphy and coworkers found that this ratio influences

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Table IV. Differences of some of the factors in different years of the study.

Year	Mortality rate (%)	Mean age (year)	Mean weight* (kg)	Mean RV/LV ratio	TAP* (%)
1991	10	3.6	11.25	0.59	30
1992	11.1	6.4	17	0.53	11
1993	0	7.6	19.4	0.56	50
1994	7.7	8.7	23	0.46	38
1995	16.7	6.6	16.9	0.53	27
1996	6.7	8.5	22.1	0.43	66
1997	23.1	5.8	15.9	0.55	69
1998	16.7	7.4	16.3	0.45	66
1999	13.3	10.7	29.7	0.47	60

*Significantly different between groups (p= 0.02 using single factor ANOVA test).
*Significant increase with years of operation (p= 0.03 using chi-square test).

the long-term survival as well,[15] but others didn't show a correlation between RV/LV ratio and long-term outcome.[6,17] In this study, the mean RV/LV ratio was not significantly different between non-surviving and surviving patients and we were not able to find a statistically significant cut off point for increasing mortality risk for this index. However, by grouping the patients according to their RV/LV ratio, we will see an increasing incidence in mortality as this index increases (Fig. 1).

Although some reports have shown increase in operative mortality in patients requiring TAP,[4,9,10,22] in a review of 208 children who had repair of tetralogy of Fallot, there was no association between operative mortality and the use of a TAP.[14] It is not certain whether higher mortality is a result of more severe RVOTO or it is an independent risk factor. In spite of a nearly insignificant difference in overall mortality between the TAP and non-TAP group in our patients, looking at the primary and the two-stage groups separately, we see a big difference in mortality between TAP and non-TAP in the primary but not the two-stage group.

The lower incidence of TAP and higher mortality in the primary group may bring this suspicion that if this group had been corrected with more radical reconstruction of RVOT (i.e. with TAP), the mortality would have been less. If the mortality and the RV/LV ratio both were higher in the primary group, we could conclude that more TAP's were required in the primary group. But having an equal ratio of RV/LV in all groups including non-surviving and surviving primary repairs without TAP indicates that they could not

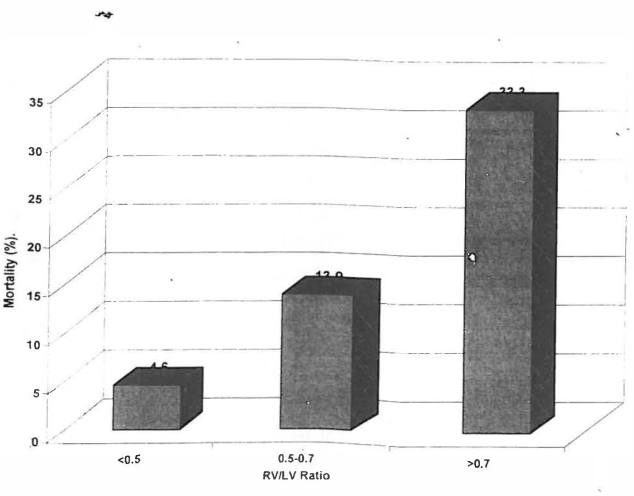

Fig. 1. Mortality rate in groups with different RV/LV ratio.

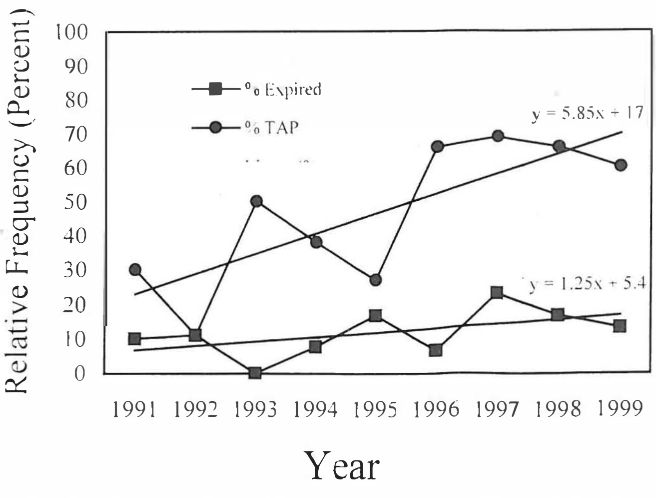

Fig. 2. Changes of mortality and percentage of the patients that needed TAP.

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benefit from TAP. In other words, because the mortality is higher and the RV/LV ratio almost the same, we may conclude that those patients in the primary group had more unbalanced ventricles as a cause of increased mortality. To justify this, we believe that shunting will prepare the left ventricle for accepting the extra blood volume that will reach the left ventricle after total correction.

There has been improvement in early mortality after primary repair of tetralogy of Fallot and more recent studies show lower early postoperative mortality than two-stage repair.[12,16,21] Although we have a lower mortality in our two-stage group and we have only a 2.5 percent procedural mortality for our shunts, since we have not performed total correction in all shunted cases, there remains a question of mortality while waiting for total correction.

There was a significantly higher incidence of TAP in the two-stage group. This may support the statement regarding annular growth regression with shunting. However, having older patients in the two-stage group without TAP may indicate that shunting has indeed induced annular growth. The finding that the RVOT-P group had a statistically significant longer time interval between shunt and total correction than the TAP group is another support for annular growth with shunt. We believe the higher incidence of TAP in the two-stage group mostly indicates that we have shunted tetralogies with more severe RVOTO.

The mortality rate slightly increased along the time of study. The relation between mortality and TAP along with increase in the percentage of patients that needed TAP with the years of study indicates that this increase in the mortality rate mostly reflects operating more severe cases with time. The mortality rate has increased with a smaller slope than the TAP rate (Fig. 2), which may be attributed to the surgeon's learning curve.

CONCLUSION

Our patients generally do better on two-stage repair, because of their age. The long period of low pulmonary blood flow has induced unbalanced ventricles for them and exaggerated RVOTO due to muscle hypertrophy. In such patients, shunting will prepare the left ventricle for accepting the extra blood volume that will reach the left ventricle after total correction. Nevertheless, individualization of each case is mandatory and selecting special indices such as left ventricular compliance, to decide accordingly and categorize the patients to one or two-stage operations needs further investigation.

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