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Neonatal repair of tetralogy of Fallot results in improved pulmonary artery development without increased need for reintervention☆

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Abstract

Objective: Despite a continuous improvement in operative outcome the optimal timing for the repair of tetralogy of Fallot (TOF) remains controversial. The purpose of this study was to evaluate the results following neonatal repair of TOF and the need for reintervention associated with this strategy. **Methods**: Retrospective review of 66 consecutive patients with TOF and confluent pulmonary arteries, who underwent repair immediately after diagnosis. Group I ($n=46$) had a median age of 5 days (1–29) and Group II ($n=20$) had a median age of 56 days (32–270). A transventricular approach was used in all cases, and 58/66 (88%) patients received a transannular patch. The median follow-up interval was 35 months (1–79). **Results**: There were three early deaths (4.5%) and no late deaths for an actuarial survival rate of 95.4% at 1 and 5 years. Univariate analysis of patient and procedural variables demonstrated that early mortality was only influenced by associated non-cardiac conditions ($P=0.04$). At median interval of 9.8 months (3–41), 12 patients required additional intervention. During the follow-up period, a significant increase in Nakata index was observed only among neonates. Freedom from reintervention at 1 month, 1 and 5 years was: 100, 84.2 and 81% in group I and 100, 84, 78.9% in group II. Surgical weight below 2.5 kg ($P<0.001$), low arterial saturation in the early postoperative period ($P=0.04$) and small preoperative branch pulmonary artery size (<0.01) were associated with need for reintervention during follow-up. **Conclusions**: Elective repair of TOF in neonates with confluent central pulmonary arteries has excellent results in the absence of significant associated non-cardiac conditions. While enhancing the development and growth of the pulmonary arteries, neonatal repair affords a freedom from reintervention no different from patients repaired during infancy. Preoperative weight <2.5 kg and small left pulmonary artery size are associated with higher incidence reintervention during follow-up.

Keywords: Cardiac surgery; Tetralogy of Fallot; Neonatal repair

1. Introduction

Although it has been five decades since the original report on the surgical repair of tetralogy of Fallot [1], the optimal management strategy and its timing remain a matter of debate. In the current era, primary surgical correction in infants continues to gain increasing acceptance [2–4]; however, initial palliation followed by repair still remains favored by some [5]. In the early 1980s, Castaneda et al. provided a compelling argument supporting a strategy of early repair. This included minimizing the effects of hypoxia, optimizing ventricular function, reducing the incidence of arrhythmias and allowing normal development of the heart and other organs [3,4,6–8]. This philosophy, coupled with the recent improvement in the outcomes of newborns undergoing surgical correction for other congenital heart defects, [9–11] has led to the repair of TOF in neonates in some centers [4,6,12]. Although this practice has met with encouraging early results, its later impact on pulmonary artery development and need for reintervention remain unknown.

The purpose of this study was to evaluate the early and intermediate outcomes following repair of TOF in the neonatal period, with particular emphasis on pulmonary artery growth and the need for reintervention associated with this strategy.

2. Material and methods

A search of the Nemours Cardiac Center database identified 66 consecutive patients with TOF and confluent pulmonary arteries, who underwent repair at the Alfred I. DuPont Hospital between January 1998 and March 2004. The usual management strategy was early repair immediately after diagnosis, regardless of symptoms. However, some patients underwent repair beyond the neonatal period when diagnosis or referral was delayed. Patients with non-confluent pulmonary arteries, absent pulmonary valve

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syndrome, associated A-V canal defect or pulmonary blood flow predominantly from systemic to pulmonary vessels were not included in this review.

Data was obtained from hospital records, operative and catheterization reports, outpatient visits and referring physician notes.

Cardiac diagnosis was based on transthoracic echocardiography using subcostal, parasternal, apical four chamber and suprasternal views in all cases. Cardiac catheterization was used only occasionally to further elucidate the anatomy of the native pulmonary arteries and to define the presence of large systemic to pulmonary connections.

All repairs were performed through median sternotomy using cardiopulmonary bypass with a period of deep hypothermic circulatory arrest, and a single dose of crystalloid cardioplegia. Cardiopulmonary bypass time averaged 65.59±25.4 min, with a mean deep hypothermic circulatory arrest time of 37.79±11.4 min. The ventricular septal defect was closed with a Dacron patch via transventricular approach in all children. Right ventricular outflow tract reconstruction was performed using a transannular patch, which extended onto the proximal left pulmonary artery in 58 patients (88%). Of the eight patients who did not receive a transannular patch, six of them had pulmonary valve annulus with a mean *Z*-score greater than −2. In two patients, who had a large coronary artery crossing the RVOT, the right ventricular outflow reconstruction was performed using a left atrial autograft flap [13]. If a patent foramen ovale was present this was left open in all patients. The sternum was routinely closed and a short-acting narcotic infusion was utilized with the aim of early extubation. The follow-up data regarding survival and reinterventions was complete as of March 2004 for all 63 patients who survived to hospital discharge. The median follow-up for the entire cohort was 35 months (range 1-79), which was similar for both groups.

3. Statistical analysis

Standard descriptive statistic methods were used. Data are described as frequencies, median with ranges, and mean±SD, as appropriate. Data is presented with 95% confidence limits. To assess differences between groups the Mann-Whitney's test or Student's test for continuous data and Chi-square or Fisher's exact test for categorical data were used. Echocardiographic measurements concerning development of pulmonary arteries were analyzed with repeated measures ANOVA and Scheffe's post hoc test. Multivariable logistic regression analysis was performed to determine risk factors of mortality. Survival and freedom from reintervention were derived by Kaplan-Meier's method, using 95% confidence limits. Risk factors of time-related reintervention were established with Cox proportional-hazards model. A value of $P<0.05$ was considered statistically significant.

4. Results

There were 46 neonates (Group I) and 20 infants (Group II). Five patients (7.5%) had pulmonary atresia and 16 patients (24.2%) had associated cardiac diagnoses (Appendix A). Nine patients (13.6%) had an additional non-cardiac diagnosis or genetic syndromes. A deletion of 22q11 region was detected in four and Trisomy 21 in two patients. One patient had a pentalogy of Cantrell and another had VACTERL syndrome. One patient had left diaphragmatic hernia associated with left lung hypoplasia. A total of six patients (three in each group) had documented hypercyanotic 'spells'. Eleven patients received mechanical ventilation and 12 were receiving a prostaglandin E1 infusion in the neonatal group. A similar proportion of symptomatic patients was observed in each group (14/46 neonates vs. 6/20 non-neonates).

Table 1
Group characteristics

Variable	Total ($N=66$)	Neonates ($N=46$)	Nonneonates ($N=20$)	P
Birth weight (kg)	3.01±0.6	3.01±0.5	2.96±0.9	0.96
Surgery weight (kg)	3.45±1.0	3.10±0.6	4.29±1.3	<0.001
EGA (weeks)	37.59±5.8	38.54±1.8	38.0±3.5	0.5
Age (days)	31.43±50.2	8.41±8.02	84.4±65.26	<0.001
DHCA (min)	37.79±11.4	36.89±10.1	39.85±13.9	0.59
CPBT (min)	65.59±25.4	62.32±19.3	73.1±35.2	0.45

EGA, gestational age; DHCA, deep hypothermia circulatory arrest time; CPBT, cardiopulmonary bypass time.

Patient characteristics in Table 1 show a significant difference for age ($P<0.001$) and weight at the time of surgery ($P<0.001$), but no differences concerning gestational age, cardiopulmonary bypass and circulatory arrest times between groups.

There were three hospital deaths (4.5%), all symptomatic patients. A female infant with confluent pulmonary arteries and two large systemic to pulmonary collaterals transferred from another institution who underwent tetralogy repair including unifocalization of two large collaterals with the right ventricular outflow tract. This patient expired on the 28th postoperative day secondary to low cardiac output and sepsis. The second death occurred in a newborn with left diaphragmatic hernia and hypoplastic left lung who underwent dehernia followed by TOF repair 3 days later. The postoperative course was complicated by severe pulmonary artery hypertension, which despite nitric oxide and extracorporeal circulatory support led to his death on the third postoperative day. The third death occurred on a 2.4-kg preemie with VACTERL syndrome who developed necrotizing

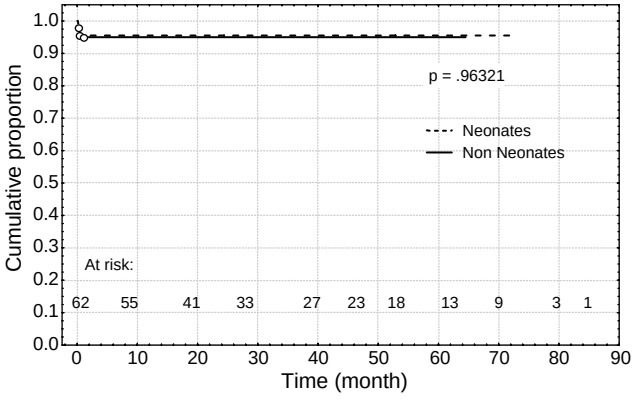

Fig. 1. Actuarial survival curve.

Table 2
Changes of the pulmonary artery Z-score and Nakata index

	Preoperative (1)	P (1 vs. 3)	Discharge (2)	P (1 vs. 2)	Follow-up (3)	P (2 vs. 3)
RPA Z-score; median (range)						
Neonates	−0.87 (−3.5-0.98)	0.001	−0.87 (−3.18-1.18)	0.99	−0.63 (−1.4-1.69)	0.001
Non-neonates	−1.59 (−2.4-1.4)	0.31	−1.14 (−2.7-0.03)	1.0	−0.91 (−2.3-0.18)	0.28
P (neonates vs. non-neonates)	0.98		0.99		0.99	
LPA Z-score; median (range)						
Neonates	−1.41 (−3.7-1.23)	<0.001	−1.30 (−3.4-0.4)	0.22	−0.24 (−3.3-3.5)	0.014
Non-neonates	−1.73 (−3.2-1.3)	0.35	−1.2 (−3.2-1.73)	0.96	−0.25 (−1.8-4.6)	0.82
P (neonates vs. non-neonates)	0.98		0.99		0.74	
Nakata index						
Neonates	98.54±44.6	<0.001	107.82±45.8	0.57	159.60±55.3	<0.001
Non-neonates	119.92±68.9	0.52	118.69±46.3	0.97	157.37±58.0	0.92
P (neonates vs. non-neonates)	0.99		0.99		0.99	

RPA, right pulmonary artery; LPA, left pulmonary artery.

enterocolitis, sepsis and disseminated intravascular coagulopathy after repair. No difference in mortality was observed between groups (4.34 vs. 5.0%; $P=0.97$). Univariate analysis of patient and procedural variables demonstrated that early mortality was only influenced by the presence of an associated non-cardiac diagnosis or genetic syndrome ($P=0.02$). Multiple logistic regressions did not identify any risk factor for mortality. Due to absence of deaths among asymptomatic patients, no risk factors could be identified. There were no late deaths during follow-up in either group, for an actuarial survival rate of 95.4% at 1 and 5 years (Fig. 1).

One patient in each group received extracorporeal circulatory support. Postoperative arrhythmias occurred in 13/44 (29.5%) neonates and 4/20 (20%) infants, which was not statistically different.

The entire cohort had a median duration of mechanical ventilatory support of 5 h (range 1-122), a median postoperative intensive care unit stay of 2 days (range 1-86), and a median hospital stay of 8 days (range 3-243). No significant differences for these variables were observed between groups.

Echocardiographic determination of branch pulmonary arteries Z-scores and Nakata index was performed preoperatively and during follow-up as shown in Table 2. No significant differences concerning branch (RPA and LPA) pulmonary artery Z-scores were observed preoperatively between groups. A significant increase in both RPA and LPA Z-scores associated with a significant increase in the Nakata index was observed in the neonatal group during the follow-up period. This change did not occur immediately after surgery. A similar trend was observed among patients repaired in the non-neonatal period; however, this was not significant.

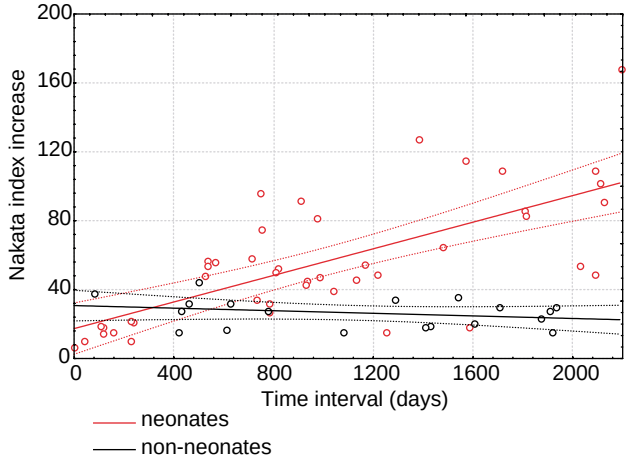

Fig. 2. Individual increase in Nakata index during follow-up (dotted lines= 95% CL).

The increase in Nakata index observed in each patient during follow-up is shown in Fig. 2. Patients repaired in the neonatal period exhibited a significant increase in Nakata index, in contrast to patients repaired beyond the neonatal period who demonstrated smaller morphometric changes resulting in a flattening of the growth curve.

Additionally, although the operative relief of right ventricular outflow obstruction was effective as demonstrated by a significant drop in the mean preoperative gradient estimated by echocardiogram at the time of discharge (54.6±18.5-18.4±8 neonates and 60.2±21.4-12.8±5.2 infants; $P<0.05$), a modest yet non-significant increase in the outflow tract gradient was observed in the entire cohort during follow-up.

Twelve (19.04%) patients underwent a total of six operative and six cardiac catheterization procedures at a median interval of 9.8 months (range 3-41 months) after repair. The indications for these interventions are shown in Table 3. The interventions on the RVOT were equally distributed between groups, and all interventions on the left pulmonary artery occurred in patients repaired in the neonatal period. Six patients who previously had received a transannular homograft patch underwent additional muscle

Table 3
Reinterventions during the follow-up period

Variable	Total (N=63)	Neonates (N=44)	Non-neonates (N=19)
Total, *n* (%)	12 (19.0)	8 (18.2)	4 (21.0)
Surgery RVOTO	6 (9.5)	3 (6.8)	3 (15.7)
Catheterization			
LPA stenosis	5 (7.9)	5 (11.3)	0 (0.0)
RPA stenosis	1 (1.6)	0 (0)	1 (5.2)

RVOT, right ventricular outflow tract; LPA, left pulmonary artery; RPA, right pulmonary artery.

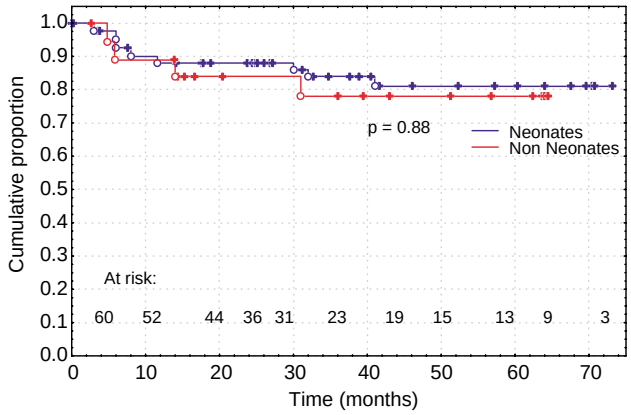

Fig. 3. Freedom from all reinterventions (surgical and catheterization).

division or excision and RVOT homograft patch augmentation using cardiopulmonary bypass. Concomitant closure of patent foramen ovale (2) and residual ventricular septal defect (2) was performed in four patients. The other six patients had a balloon angioplasty and stent placement to relieve branch pulmonary artery stenosis.

The 1-month, 1 and 5-year freedom from reintervention (surgical or catheterization) were: 100, 84.2 and 81% for neonates and 100, 84 and 78.9% for infants (Fig. 3). No difference in reintervention rate was observed between groups ($P=0.88$). Univariate analysis identified surgical weight under 2.5 kg ($P=0.003$), smaller preoperative left pulmonary artery diameter ($P=0.01$), and lower arterial saturation at 1 h ($P<0.001$), 6 h ($P=0.012$) and 12 h post operatively ($P=0.002$) to be associated with reintervention during follow-up. Multivariate analysis only identified operative weight ($P=0.02$) and lower arterial blood saturation in first postoperative hour ($P=0.04$) as predictors of reintervention (Table 4.).

5. Discussion

The surgical management of tetralogy of Fallot has evolved over time from a two-stage approach into a strategy of primary surgical correction in infancy. More than two decades ago Castaneda and colleagues presented a rationale for the early anatomic and physiologic correction of TOF, which included the elimination of the stimulus for hypoxia, ventricular hypertrophy, and the preservation of myocardial function [3]. This philosophy coupled with the advent of prenatal diagnosis and the remarkable improvement in the operative outcome of newborns undergoing surgical management for other forms of congenital heart disease in the current era [9–11] have contributed to support the practice of complete repair of TOF in the newborn period [4,6,12]. Our report summarizes a single-institution experience with early repair of TOF and demonstrates that complete repair in the newborn period enhances pulmonary artery growth and is associated with low mortality.

Despite the obvious differences for age and weight at the time of surgery, no difference in mortality was observed between groups. This is consistent with previous reports on the early repair of symptomatic and asymptomatic patients with TOF. [4,6,12] In contradistinction to some of those reports our analysis revealed that low surgical weight was not associated with hospital mortality. It was the presence of associated non-cardiac co-morbidities which the univariate analysis showed to have a negative influence on hospital survival. This association has also proven to be a risk factor for poor outcome following other forms of complex neonatal surgery, and reflects our limited success neutralizing these high-risk conditions. Despite significant institutional expertise and technological advances, major existing co-morbidities, continue to pose a significant challenge in the perioperative care of these patients [14].

Recently, a large retrospective study found that age less than 3 months at the time of TOF repair was associated with longer time to normalization of serum lactate, longer time to extubation, and increased length of hospital stay. However, mortality was only associated with older age at the time of repair [15], suggesting the possibility that younger patients may have a better tolerance for the repair.

The fact that our data did not demonstrate any differences in the duration total circulatory support time (cardiopulmonary bypass plus hypothermic circulatory arrest), mechanical ventilatory support, intensive care stay and total length of hospital stay between groups reflects the feasibility of neonatal repair and the consistent approach used in the perioperative care of this cohort.

All deaths in our study occurred in symptomatic patients. This observation confirms the notion that early repair of TOF in asymptomatic neonates is safe and these patients were not exposed to additional risk because of the strategy utilized.

Analysis of central pulmonary arteries development showed that despite the significant differences in preoperative age and weight between neonates and infants, the pulmonary arteries diameters, *Z*-scores and Nakata index were not significantly different between groups, suggesting that in the presence of significant right ventricular outflow obstruction the pulmonary arteries had not grown, which

Table 4
Factors influencing reintervention during follow-up

Variable	Reinterventions ($N=12$)	No reinterventions ($N=54$)	*P* univariate	*P* multivariate
WOS (kg)	2.71±0.38	3.62±1.04	<0.001	0.02
WOS<2.5 kg; *n* (%)	6 (50)	7 (12.9)	0.003	-
LPA (mm)	3.22±0.5	4.21±1.9	0.001	-
Sat 1 h postoperatively	86.20±4.8	94.46±8.2	<0.001	0.04
Sat 6 h postoperatively	84.70±6.9	90.85±7.8	0.012	-
Sat 12 h postoperatively	83.70±5.3	90.89±8.9	0.002	-

WOS, weight of surgery; LPA, left pulmonary artery; Sat, arterial oxygen saturation.

confirms the observations reported by Geva et al. [16]. In his detailed morphometric study, patients who underwent surgical repair in the first year of life showed no significant changes in the indexed diameters of the branch pulmonary arteries during the study period.

Provision of normal pulmonary flow in the neonatal period has been shown to play an important role fostering the development of pulmonary vasculature and alveologenesis [17]. Our observations included a substantial change in the branch pulmonary arteries diameter and *Z*-score not immediately after surgical correction, but during the follow-up period. The preoperative values for branch pulmonary artery *Z*-score and Nakata index which were substantially low, increased in both groups; however, this change was only significant among neonates. It is possible these observations are due to a greater growth potential of the pulmonary arteries following correction of TOF in the neonatal period.

Although the primary surgical correction of TOF in infants and neonates has gained increasing acceptance in the current era, the lack of data concerning the intermediate outcome and the need for reintervention makes the true value of this approach difficult to ascertain. Our data provides further evidence regarding the applicability and safety of this approach and its influence on pulmonary artery development.

The reported incidence of reinterventions following repair of TOF ranges between 15 and 40% [18,19–21]. Factors contributing to recurrent outflow obstruction or pulmonary artery restenosis include restrictive growth of the RVOT, early residual stenosis, angulation or stenosis of the left pulmonary artery, extension of the ductal tissue into the origin of the left pulmonary artery; the bifurcation of the pulmonary artery being reported as the most frequent site of restenosis in patients operated beyond the neonatal period [22,23].

Over a median follow-up of 35 months 12 patients required a reintervention due to either right ventricular outflow obstruction or branch pulmonary artery stenosis. The predominant site of obstruction was the right ventricular outflow, which had a similar distribution between groups. However, left pulmonary artery stenosis was more common in patients who underwent neonatal repair.

Despite the common use of a transannular patch in a large proportion of our patients, stenosis at the right ventricular outflow level is likely related to the tendency to minimize the ventriculotomy in order to reduce the potential for right ventricular dysfunction. As reported by Bacha et al., the placement of a transannular outflow patch during the repair of TOF in infancy is associated with lower incidence of right ventricular outflow tract obstruction [24]. This is in contrast to our observation where all patients who presented with recurrent outflow obstruction had in fact received a transannular patch, implicating the restrictive nature of a very conservative ventriculotomy as the culprit for the residual obstruction.

Regarding left pulmonary artery (LPA) restenosis, univariate and multivariate analysis found that preoperative weight, and small LPA size (in mm) were associated with a higher likelihood of reintervention. In our cohort, all patients with low operative weight had a low birth weight for gestational age. This has been previously shown to be a factor influencing growth potential independently [25], therefore the higher likelihood of inadequate pulmonary artery growth after surgical repair.

It is known that preoperative branch pulmonary arteries size alone cannot be a good predictor of postoperative size in the presence of significant outflow obstruction. Therefore, it is imperative that right ventricular outflow tract obstruction be relieved in order to increase the absolute diameter of the compliant arteries and to decrease the gradient across RVOT.

Additionally, patients with low arterial saturation within the first hours of the postoperative period were at significant risk for reintervention. In our group, the low saturation during the initial postoperative hours did not correlate with preoperative pulmonary arteries size and most likely was related to some degree of restrictive right ventricular physiology. Although the multivariate Cox regression did not demonstrate the left pulmonary artery diameter or *Z*-score as a risk factor for reintervention, five patients in the neonatal group received a stent into the left pulmonary artery.

The 1-month, 1 and 5-year freedom from reintervention (either surgical or catheterization) were: 100, 84.2 and 81% for neonates and 100, 84, 78.9% for infants. This relative improvement on the freedom from reintervention compared to other series could be attributed to the small proportion of patients with pulmonary atresia and the exclusion of patients without confluent central pulmonary arteries from this cohort.

When considering the potential need for reintervention related to the presence of long-standing pulmonary insufficiency, it is possible that delaying the age of repair among asymptomatic patients could reduce the use of transannular patching and perhaps lessen the potential for reintervention directed at the elimination of free pulmonary insufficiency.

6. Limitations of the study

The retrospective nature of the study and the lack of randomization could make the analysis susceptible to error. The small number of deaths in the cohort and the absence of deaths among asymptomatic patients limited the analysis of potential factors influencing mortality. All hemodynamic data were based on the estimates obtained by two-dimensional echocardiogram therefore are susceptible to error.

Due to the limited follow-up, the reintervention data should be considered an approximation of the true value, which is most likely underestimated; particularly when considering the potential need for reintervention in patients who received a transannular patch. Due to the small number of reinterventions, surgical and interventional procedures have been combined, making the identification of factors associated with each specific type difficult.

In order to limit the probability of error given the number of statistical test performed we have done no more than 10 simultaneous tests and used the Bonferroni correction.

7. Conclusions

Elective repair of TOF in neonates with confluent central pulmonary arteries has excellent results in the absence of associated non-cardiac conditions. While enhancing the development and growth of the pulmonary arteries, neonatal repair affords a freedom from reintervention no different from patients repaired during infancy. Preoperative weight <2.5 kg and small left pulmonary artery are associated with higher incidence reintervention during follow-up. The use of a transannular patch maybe associated with an increase in the number of reintervention over time.

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Appendix A

Left superior vena cava—9 patients
Anomalies of coronary arteries—5 patients
Major aorto-pulmonary collaterals—5 patients
Right aortic arch with mirror image branch—3 patients
Atrial septal defect secundum type—1 patient
Partial anomalous pulmonary venous connection—1 patient
Retroaortic innominate vein—1 patient

Appendix B. Conference discussion

Dr G. Sarris (*Athens, Greece*): Our center in Athens, the Onassis Center, has employed the policy of repairing tetralogy at about a year of age with selective use of shunting if severe symptoms occur earlier. Using this policy, over the last 7 years we have repaired a series of approximately 150 consecutive patients, for which total mortality (including for those patients who required shunting in the very first few months of life) is zero. Late reoperation rate is limited to one patient so far.

The surgical technique employed was transatrial/transpulmonary. Others have also employed transatrial/transpulmonary repair with this management protocol, notably Dr Mee in Melbourne and also at the Cleveland Clinic, and others have used transatrial repair earlier in life, down to the neonatal period, and I believe have reported similar results.

My question to you is, considering that you're reporting here an almost 5% mortality and a substantial reoperation rate, are you considering altering your approach as far as either timing of surgery or as far as the method of operation to be applied, particularly with reference to possibly using the transatrial approach?

Dr Kolcz: With reference to mortality, all patients who died in these series had severe associated non-cardiac diagnosis, for example, pentalogy of Cantrell or left diaphragmatic hernia.

Well, is it possible that if these patients had been shunted that probably would have been a better choice for them?

In general, the approach of our institution is to correct the defect in the neonatal period.

The freedom from reintervention rate is comparatively low. It's something about 80% at 5 years. Taking into account that the rather conservative approach to relive the RVOT obstruction was used, these findings are good and related to doing a small incision within the infundibulum which should be large enough to close the ventricular septal defect and small enough to avoid dysfunction of the right ventricle.