

Intermediate results with correction of tetralogy of Fallot with absent pulmonary valve using a new approach[☆]

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Abstract

Objective: The intermediate results achieved with a new technique for primary repair of tetralogy of Fallot with absent pulmonary valve syndrome are reported. **Method:** Apart from correction of tetralogy of Fallot, this approach includes translocation of the pulmonary artery anterior to the aorta and away from the tracheobronchial tree. Since November 1998 this technique has been employed in three symptomatic newborns and three infants with a diagnosis of tetralogy of Fallot with absent pulmonary valve. The median age at surgery was 37 days (range 14–256 days). The median weight at operation was 3.4 kg (range 2.9–4 kg). All patients had severe respiratory problems and congestive heart failure with cyanosis. **Results:** There was no early or late death during the follow-up (median 27 months). One patient required redo due to failure to thrive as a consequence of right ventricle volume overload. Valved conduit was inserted to pulmonary position. Respiratory symptoms disappeared or were significantly reduced in all patients. Postoperative computed tomographic scan showed no compression of trachea and main bronchi, pulmonary artery was away from tracheobronchial tree in all patients. Patients are doing well with adequate growth. **Conclusions:** The new technique described here has a potential to reduce or eliminate bronchial compression by pulmonary artery. Translocation of pulmonary artery anterior to the aorta takes the dilated pulmonary artery away from the trachea and bronchial tree. This approach was found to be technically feasible and can be useful especially in symptomatic newborns and infants. © 2002 Elsevier Science B.V. All rights reserved.

Keywords: Tetralogy of Fallot; Absent pulmonary valve syndrome; Respiratory distress

1. Introduction

The absent pulmonary valve syndrome occurs in 3–6% of patients with tetralogy of Fallot (TOF). This syndrome is physiologically distinctive from other form of TOF because of tracheobronchial compression resulting from massive dilatation of the main pulmonary artery (PA) and its first- and second-order branches, and from the abnormal branching of segmental arteries. Clinical presentation reflects the degree of respiratory distress secondary to airway obstruction and infections and heart failure as a result of left to right shunt [1,2].

Since 1998 we have used a new technique that should eliminate or reduce compression of tracheobronchial tree.

Apart from correction of TOF, this approach includes translocation of the PA anterior to the aorta and away from the airways [3].

The aim of this study is to report the intermediate results achieved with this approach with all consecutive patients undergoing primary repair in our center.

2. Materials and methods

2.1. Patients

Six patients with tetralogy of Fallot with absent pulmonary valve (TOF/APV) were operated on between November 1998 and December 2000 at the Children's Hospital of Bratislava (Table 1). Three patients underwent surgery during infancy and three underwent neonatal repair (age < 30 days). Median age at surgery was 37 days (range 14–256 days). Median weight at operation was 3.4 kg (range 2.9–4 kg).

All patients had severe respiratory problems and conges-

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Table 1
Data on patients^a

Patient no.	Age (days)	Weight (kg)	Symptoms	Homograft to PA position	Extubated (days)	Outcome
1	254	4.0	Repeated chest infections, FTT	No	7	Well, infrequent chest infection
2	109	3.3	Repeated chest infections, FTT	No	2	Well, no respiratory symptoms
3	27	3.1	Respiratory distress, heart failure, cyanosis	No	7	Well, infrequent chest infection
4	24	2.9	Respiratory distress, heart failure, cyanosis	Yes	13	Well, no respiratory symptoms
5	14	3.6	Respiratory distress, heart failure, cyanosis	Yes	7	Well, no respiratory symptoms
6	48	3.5	Repeated chest infections, heart failure	Yes (monocusp)	4	Well, infrequent chest infection

^a PA, pulmonary artery; FTT, failure to thrive.

tive heart failure with cyanosis. Surgery in neonates was performed on semi-urgent basis due to serious respiratory compromise. Persistent bronchopulmonary infection requiring prolonged and repeated hospital admission and failure to thrive were the main indication for surgery in infants. No patient required either assisted ventilation or extracorporeal membrane oxygenation before surgery.

Associated intracardiac anomalies other than TOF and ostium secundum defect were present in three patients, including right aortic arch ($n = 2$) and multiple aortopulmonary collaterals ($n = 1$) which required preoperative coil embolization.

2.2. Surgical technique

All patients underwent complete repair incorporating the standard intracardiac portion of TOF repair (resection of right ventricular tract (RVOT) obstruction, patch closure of the ventricular septal defect (VSD)) and translocation of the pulmonary artery anterior to the aorta. The idea of operation is to bring the PA anterior to the aorta and away from tracheobronchial tree.

This approach anticipates wide deliberation of the ascending aorta, aortic arch and brachiocephalic vessels. The superior vena cava (SVC) is dissected free and the azygos vein is transected to improve mobility of SVC. The left and right pulmonary arteries, including the first pulmonary artery branches in the hilum of each lung, are dissected free and mobilized. After TOF repair a transverse aortotomy is done above the commissure and tubular or triangular segment of the aorta is resected (Fig. 1). This maneuver brings the future ascending aorta down and to the left. The pulmonary artery is transected above the annulus and brought anterior to the aorta. At this point end-to-end anastomosis of ascending aorta is performed. Finally, a direct connection between pulmonary artery and RVOT is accomplished (Fig. 2). At this moment, anterior/posterior PA plication or homograft insertion can be done, if necessary. Details of this approach were published elsewhere [3].

The RVOT was reconstructed with valved homograft in two newborns; a monocusp valve was utilized in one infant. Combined anterior and posterior plication of pulmonary artery was performed in three patients.

2.3. Data analysis

Data are presented as median and range.

2.4. Functional outcome and follow-up

Regular clinical check-up was performed at 3-month intervals during the first postoperative years and at 6–12-month intervals later on. All patients underwent two-dimensional echocardiography (ECHO) and Doppler assessment of the intracardiac repair as well as computed tomography (CT) of the mediastinum three months postoperatively.

Follow-up data were obtained in all patients. The median follow-up for the whole group is 27 months (range 17–35 months).

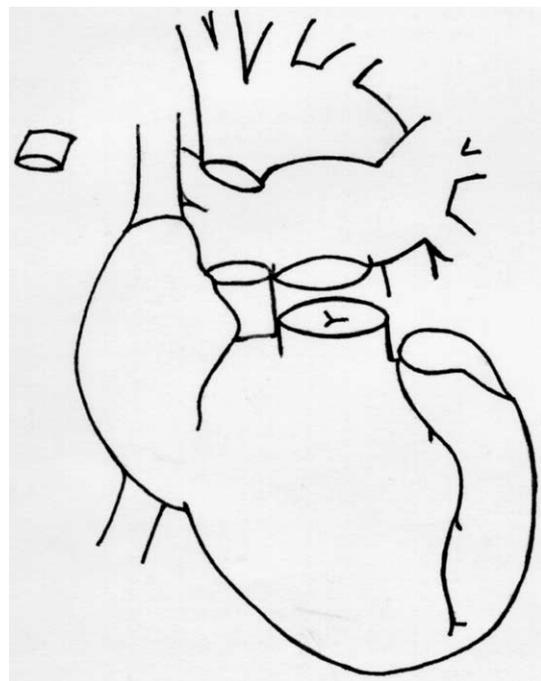


Fig. 1. Triangular segment of the aorta is resected. The pulmonary artery is transected above the annulus.

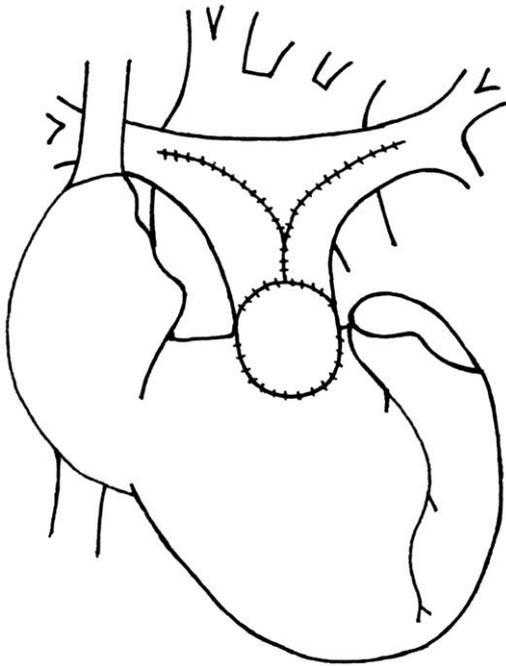


Fig. 2. The pulmonary artery is brought anterior to the aorta. The direct connection between pulmonary artery and right ventricle outflow tract, or homograft insertion is accomplished.

3. Results

3.1. Early results

There was no early death. Hemodynamically, the postoperative course was generally uneventful; one newborn had delayed sternal closure and inotropic requirements varied from 1 to 4 days. Transient right ventricular failure with pleural effusion was noted in three patients.

The duration of mechanical ventilation varied from 2 to 13 days (median 7 days) and the intensive care unit (ICU) stay varied from 9 to 25 days (median 12 days). Recovery from respiratory distress was delay in one newborn due to inhalation pneumonia requiring mechanical ventilation for 13 days.

3.2. Late results

There was no late death. Three months after neonatal repair one patient required reoperation for failure to thrive. Valved conduit was inserted into the pulmonary position. His postoperative course was uneventful. He was the only primarily operated newborn in whom the valve homograft was not utilized for reconstruction of RVOT. Another patient had ventriculo-peritoneal shunt implanted due to hydrocephalus.

On Doppler echocardiography two patients with homograft had gradients of 30 and 17 mmHg, respectively. Small residual VSD was noted in one patient. There were no signs of SVC obstruction.

At follow-up, respiratory symptoms completely disappeared (three patients) or were significantly reduced. Post-



Fig. 3. Postoperative CT scans 3 months after operation. There is no compression of main bronchi; pulmonary artery is translocated anterior to the aorta.

operative CT scan showed no compression of trachea and main bronchi, pulmonary artery was away from tracheo-bronchial tree in all patients (Fig. 3). In the hilar portion of the lungs of patients without plication of pulmonary arteries, there was an immediate contact between dilated pulmonary artery and its branches and mainstem and segmental bronchi (Fig. 4). These patients had slight dystelectasis, especially in the right upper lobe of the lung.

Neurological examination revealed only mild tone alteration (hypotonia) in one patient at 1 year of age; the rest of patients showed normal neurological development.

The patients are doing well and are growing adequately. All patients are on medication (Digoxin and/or diuretics) but no one is in need of bronchodilator therapy.

4. Discussion

Despite improvements in surgical techniques and critical

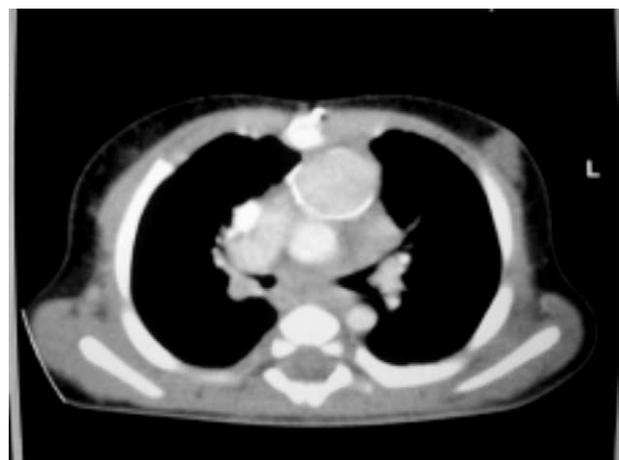


Fig. 4. In the hilar portion of the lungs there is an immediate contact between dilated pulmonary artery and its branches and segmental bronchi without signs of compression.

care, controversy persists regarding the management of patients with TOF/APV. The mortality for symptomatic newborns and infants remains considerable secondary to airway obstruction by dilated PA [2,4–6].

Massively dilated right and left PA up to the hilum are amenable to surgical intervention [2–6]. Unfortunately abnormalities of arborization, with tufts of arteries encircling and compressing the intrapulmonary bronchi, cannot be addressed during the surgery [1]. This can partially explain the high rate of failure with the treatment of the youngest, symptomatic group of patients.

A number of surgical techniques for reduction of bronchial obstruction have been proposed with questionable results. The method of choice especially in symptomatic newborns and infants is still controversial. All strategies have focused on plication, reduction of the anterior or posterior wall of the normally positioned PA with or without pulmonary valve replacement [4–6].

Our approach applies a well-known technique (Lecompte maneuver) [7] that has the potential to reduce or eliminate bronchial compression by PA. Translocation of PA anterior to the aorta displaces the dilated PA anterior, away from the trachea and bronchial tree. There are several technical pitfalls to keep in mind during the performance of this procedure. It is essential to gain adequate room between SVC and ascending aorta for the translocated right PA. In addition to SVC mobilization, shortening of the ascending aorta allows the aorta to ultimately reside posteriorly and to the left of its usual location. This maneuver calls for a thorough mobilization of the aortic arch and brachiocephalic vessels. Shortening of the ascending aorta and mobilization of PA beyond pericardial reflection avoids the potential compression of right coronary artery and SVC. Another relevant detail is shortening of the left PA (which is always too long) by oblique transection of the PA trunk with connection to the RVOT. Finally, insertion of valve homograft with anterior and posterior plication of PA can decrease wall tension and prevent later development of aneurysmal dilatation of PA [3]. This is our current approach for symptomatic newborns and infants, which was found to be technically feasible and useful. Our management strategy is consistent with the report of McDonnell and co-workers who recommended not only anterior and posterior plication of PA but insertion of homograft, especially in newborns and infants [6].

There is no mutual consensus on the timing for surgery for babies presenting with TOF/APV. Obviously, symptomatic patients need to proceed directly to surgery. In addition to this, we believe that early repair in asymptomatic patients can eliminate the potentially harmful effect of dilated PA on the tracheobronchial tree.

In conclusion, from 1998 we started to use a new technique that has the potential to eliminate or reduce bronchial compression by PA. Apart from correction of TOF, this approach includes translocation of the PA anterior to the aorta and away from the airways, and insertion of valve

homograft with anterior and posterior plication of PA. With the technical aspects of this procedure well accomplished, functional results are quite encouraging with zero mortality events in the youngest age group of patients. The small number of patients and the short period of follow-up preclude any firm conclusions.

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Appendix A. Conference discussion

Mr J. Monro (Southampton, UK): I think you showed you had to go back and insert a homograft in one patient. I must say, we routinely use a homograft in all these neonates and young infants with this condition. What I have done is to put a Hegar dilator down the pulmonary arteries and oversew it to narrow it to whatever diameter you want, taking great care not to mobilize the back of the pulmonary artery, and then leave the stitch that you have used and at the end of the procedure, bring it up through the chest wall and just tie it. So this hitches the pulmonary artery forward, and certainly that opens up the bronchi, which is the main problem with this disease.

Dr Hraška: Of course, it is possible to perform the operation like this, and the results can be very good.

Dr J. Amato (Chicago, IL, USA): I just wondered, how much of the aorta did you remove and did you have any problem with the LeCompte maneuver in coming forward, and if you did, in one of the earlier papers today I heard that they used a piece of that aorta to serve as a junction or a bridge between the pulmonary artery and the right ventricle?

Dr Hraška: Well, actually we haven't had a problem to bring the pulmonary artery anterior because the operation was performed in very small babies. So you usually don't have this problem in very small babies. In fact, one has to be very careful about the resection of the aorta because if you resect too much, you can be very close, especially to the origin of the left bronchus. It depends, of course, on the aortic arch anatomy. So one has to be very careful about it.

Dr Amato: I guess what I mean is, do you do a circumferential resection or just an anterior, a piece of the anterior wall to pull it forward?

Dr Hraška: It is like a tubular, circumferential resection.

Dr B. Asfour (Münster, Germany): I have considered to use your technique because you have published it before; however to my understanding, many of these children have bronchomalacia. So I'd rather not want to dissect the pulmonary artery away from the bronchus, and then by reducing the size of the pulmonary arteries there is some tension on the connection and the adhesion of the pulmonary artery which then keeps the bronchus open because it is so soft. Therefore we used bronchoscopy intraoperatively to monitor the bronchi after reducing the size of the pulmonary arteries open. How many of these children had bronchomalacia and did you need to use anything like stents or something to keep these bronchi open?

Dr Hraška: Well, we performed the operation in very small babies, like I mentioned, and actually we didn't check the situation as far as the bronchus by bronchoscopy. So basically I don't think that even one patient had bronchomalacia in this group. But definitely they were very symptomatic,

and I think, for the timing of the operation, if you perform the operation pretty early you can avoid all this trouble like you mentioned.

Dr Asfour: I refer to the same-age very young patients and bronchoscopy with these really thin fiberscopes which is possible.

Dr T. Tlaskal (Prague, Czech Republic): I would like to ask you, do you recommend to do this rather complex repair in all patients with tetralogy and a dysplastic pulmonary valve or only in the symptomatic neonates and infants? That is one question.

The other question is, I think that in some patients in the follow-up some progression of pulmonary stenosis could be expected with some postoperative dilatation of the hilar part of both pulmonary branches. Don't you think that this might be a problem in the follow-up?

Dr Hraška: Well, based on my experience, we haven't had any problem with postoperative pulmonary stenosis. As for your first question, I think this technique can be useful, especially for symptomatic patients. So there is no need to do it for everybody, I guess. But for symptomatic patients it can help because it makes sense to bring the pulmonary artery anterior and away from the tracheobronchial tree.