Cardiology in the Young (2013), **23**, 423–430
doi:10.1017/S1047951112000911

Original Article

Tetralogy of Fallot with unilateral absent pulmonary artery

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Abstract *Background:* Tetralogy of Fallot is a common congenital cardiac malformation. A rare subgroup includes unilateral absence of the pulmonary artery, either the left or the right main branch. The literature lacks an established treatment for these cases, and surgical options carry certain mortality and morbidity. *Patients and methods:* There were five patients who had single pulmonary artery and received surgical treatment among the 126 patients with the diagnosis of Tetralogy of Fallot, who were admitted to our institution between July, 2010 and November, 2011. All the patients were male. Ages ranged between 12 months and 8 years. The mean body mass index was 17.1 plus or minus 3.4 kilograms per square metre. Pulmonary artery Nakata index, Nakata index Z-score, and the McGoon index were used for the quantitative assessment of the pulmonary artery and to determine the surgical strategy. *Results:* Urgent modified Blalock–Taussig shunt operations were performed in two patients with very low oxygen saturation and haemodynamic instability. These patients are scheduled for corrective procedures on an elective basis. There was one patient who received an elective shunt procedure; however, the post-operative course was complicated with the overflow phenomenon and the patient underwent total correction with a check-valved patch used to close the ventricular septal defect. The patient required extracorporeal membrane oxygenator support in the post-operative period. There were two patients who underwent total correction of the pathology uneventfully. Mortality did not occur. Mean durations of hospital stay and follow-up were 14 plus or minus 13.4 days and 184.5 plus or minus 89.3 days, respectively. *Conclusion:* Our modest series with Tetralogy of Fallot with unilateral absent pulmonary artery indicates the feasibility of surgical correction in patients with appropriate unilateral pulmonary artery size and palliative procedures when the pulmonary artery size is smaller than that predicted for the age. Multi-centre long-term data of larger series are warranted in order to establish a treatment protocol.

Keywords: Tetralogy of Fallot; pulmonary artery; unilateral; absent

Received: 27 January 2012; Accepted: 20 May 2012; First published online: 18 July 2012

Unilateral absence of pulmonary artery is a very rare congenital cardiovascular malformation.[1–3] The pathology may briefly be defined as "single pulmonary artery", as the right or the left main pulmonary branch is atretic. Although it may occur as a solitary lesion, single pulmonary artery is usually associated with other congenital defects, the most common being Tetralogy of Fallot.[4,5]

The treatment strategies for Tetralogy of Fallot are well known, with low mortality and morbidity rates and event-free long-term survival with advanced surgical and anaesthesia techniques and medical measures in the current era. However, Tetralogy of Fallot together with single pulmonary artery is a challenging congenital cardiovascular pathology.

In this article, we present our experiences with Tetralogy of Fallot accompanied with single pulmonary artery, including surgical strategies and early and mid-term follow-up results.

Patients and methods

Between July, 2010 and November, 2011, 126 patients with the diagnosis of Tetralogy of Fallot were admitted to our institution. Of the 126 patients, there

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were five patients who had single pulmonary artery and received surgical treatment. Data of the patients were obtained from the institutional records.

The study was approved by the institutional review board. Patients were enrolled into the research after explaining the protocol in detail and following their consent.

Patient characteristics

All the patients were male. Ages ranged between 12 months and 8 years. The mean weight of the patients was 13.9 plus or minus 9.9 kilograms with a range from 10.50 to 30 kilograms and the mean height was 85 plus or minus 23.2 centimetres with a range from 65 to 115 centimetres, revealing a mean body mass index of 17.1 plus or minus 3.4 kilograms per square metre with a range from 13.75 to 22.68 kilograms per square metre. In all patients, the ipsilateral lung to the absent pulmonary artery was present. The lowest room-air oxygen saturation was 13% and the highest was 78% with a range from 13% to 78%. There were two patients who experienced hypoxic spells. All the patients had various degrees of erythrocytosis with a range from 52% to 59%; mean of 55.2 plus or minus 2.8%.

The first patient was 12 months old. He was 65 centimetres tall and weighed 6.75 kilograms, with a body mass index of 15.98 kilograms per square metre, a room-air oxygen saturation of 13%, and a haematocrit level of 55%. The left pulmonary artery was absent in the patient and the main pulmonary artery and right pulmonary artery sizes were 6 millimetres and 9.2 millimetres, respectively.

The second patient was 2 years old, weighed 5.63 kilograms, was 64 centimetres tall, and had a body mass index of 13.75 kilograms per square metre. His room-air oxygen saturation was 26%. He had a haematocrit level of 53%. The absent pulmonary artery was the left one in this case. The main pulmonary artery size was 6 millimetres and branch pulmonary artery size was 9.4 millimetres.

The third patient was 16 months old. His weight and height were 10.50 kilograms and 77 centimetres, respectively. The calculated body mass index was 17.71 kilograms per square metre. He had a room-air oxygen saturation of 65% and a haematocrit level of 57%. Among the patients, only in this case the right pulmonary artery was absent. On admittance, his main pulmonary artery was 8 millimetres and his left pulmonary artery was 11 millimetres.

The fourth patient was 4 years old, weighed 16.7 kilograms, was 104 centimetres tall, and had a body mass index of 15.44 kilograms per square metre. He had the highest oxygen saturation (78%) and lowest haematocrit level (52%) when compared with the other patients. In this patient, the left pulmonary artery was absent, with a main pulmonary artery diameter of 9.8 millimetres and right pulmonary artery diameter of 17 millimetres.

The last patient was the oldest – 8 years of age. He weighed 30 kilograms and was 104 centimetres tall. His body mass index was 15.44 kilograms per square metre. He had an absent left pulmonary artery with 65% oxygen saturation and 59% haematocrit level. The diameters of the main and right pulmonary arteries were 16 millimetres and 18 millimetres, respectively.

The demographic data of the patients including age, weight, height, body mass index, pre-operative oxygen saturation, pre-operative haematocrit level, and echocardiographic diagnosis are presented in Table 1.

Pre-operatively, all patients underwent electrocardiography, chest X-ray, transthoracic echocardiography, cardiac catheterisation, and computerised tomography. Owing to severe hypoxia and haemodynamic instability, two patients underwent operation urgently – the first two cases. The remaining three cases received cardiac catheterisation and right ventriculography, pulmonary arteriography, aortography, and pulmonary venous wedge angiography. The absent pulmonary arteries were diagnosed pre-operatively by echocardiography and confirmed with cardiac angiography and computerised tomography.

The ventricular septal defect component of the malformation was perimembraneous and of the malalignment type in all cases. In three cases, there was an additional secundum-type atrial septal defect – the first three patients. The third patient had stenotic but patent ductus arteriosus. In one

Table 1. Patients' demographics.

Patient	Age	Gender	Weight (kg)	Height (cm)	BMI	Diagnosis	sO_2 (%)	Haematocrit level (%)
1	12 months	Male	6.75	65	15.98	TOF + absent L-PA	13	55
2	2 years	Male	5.63	64	13.75	TOF + absent L-PA	26	53
3	16 months	Male	10.50	77	17.71	TOF + absent R-PA	65	57
4	4 years	Male	16.7	104	15.44	TOF + absent L-PA	78	52
5	8 years	Male	30	115	22.68	TOF + absent L-PA	65	59

cm = centimeters; kg = kilograms; L = left; PA = pulmonary artery; R = right; sO2 = oxygen saturation; TOF = Tetralogy of Fallot

https://doi.org/10.1017/S1047951112000911 Published online by Cambridge University Press

patient – the fourth case – an apical muscular small ventricular septal defect was present. There were small aortopulmonary collateral arteries in the second patient. None of the patients had coronary anomalies. Concomitant cardiac defects are presented in Table 2.

The quantitative assessment of the existing pulmonary artery and the feasibility of a corrective procedure were determined by calculating the Nakata index, pulmonary valve Z-score, main pulmonary artery Z-score, existing pulmonary artery Z-score, and the McGoon index.

Surgical procedures

All the patients underwent either a palliative or a corrective procedure, either under emergent or elective circumstances.

Palliative procedures

Palliative procedures included modified Blalock–Taussig shunts. The operations were performed through median sternotomy with cardiopulmonary bypass and normothermia. A 4-millimetre GoreTex graft was anastomosed between the brachiocephalic trunk or the left subclavian artery and the existing single pulmonary artery. Heparin was neutralised with half-dose protamine.

Table 2. Accompanying cardiovascular pathologies.

Patient	Main pathology	Concomitant pathology
1	TOF + absent L-PA	ASD
2	TOF + absent L-PA	ASD + MAPCA
3	TOF + absent R-PA	ASD + PDA
4	TOF + absent L-PA	Apical VSD
5	TOF + absent L-PA	None

ASD = atrial septal defect; L = left; MAPCA = major aortopulmonary collateral arteries; PA = pulmonary artery; PDA = patent ductus arteriosus; R = right; TOF = Tetralogy of Fallot; VSD = ventricular septal defect

Corrective procedures

Total correction was performed through median sternotomy. Aortic and bicaval cannulations were performed and the heart was arrested with antegrade cold-blood cardioplegia. Cardioprotection was achieved with repeated doses of cardioplegia every 20 minutes and hypothermia of 32°C. A left atrial vent was inserted through the foramen ovale to enhance exposure. The ventricular septal defect was closed with a Dacron patch. When needed, a check-valve system was constructed on the patch. Hypertrophied muscle bands in the right ventricle were divided. The atrial septal defect was closed primarily. The right ventricular outflow tract was reconstructed with an autologous pericardial patch extending to the existing pulmonary artery. Care was taken to spare at least one pulmonary leaflet. The pulmonary artery and the right ventricular sizes were adjusted depending on the age and body surface area of the child with the help of dilators.

Post-operative care

After the procedure, before weaning off the cardiopulmonary bypass inotropic agents, at least 5 micrograms per kilogram per minute of dopamine and, when needed, other agents such as dobutamine and adrenaline were started. All the patients were transferred to the cardiac intensive care unit while still intubated and extubated under optimal haemodynamic, metabolic, and respiratory conditions. All the patients received physiotherapy and digoxin and aspirin depending on the body weight in the post-operative period.

Results

In four patients, the left pulmonary artery (Fig 1a, b; Fig 2a, b; Fig 3; Supplementary material 1) was absent, whereas in one the right branch was missing. The data of the pulmonary artery parameters for surgical decision are presented in Table 3. The operative and post-operative data of the patients are presented in Table 4.

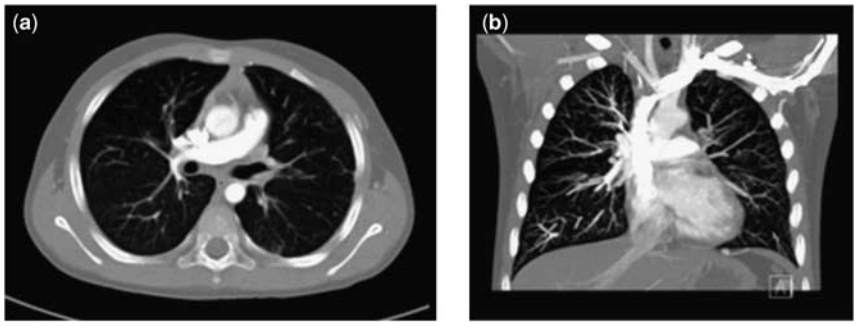

Figure 1.
*Computerised tomography of the thorax in a patient with Tetralogy of Fallot and single pulmonary artery (**a**, axial view; **b**, sagittal view).*

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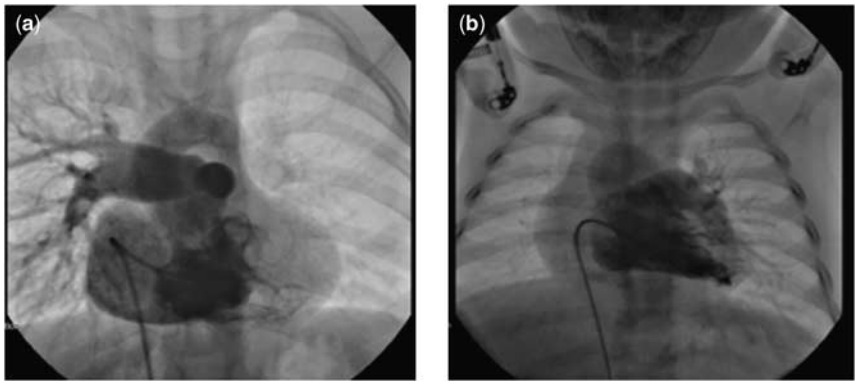

Figure 2.
*Angiography of the pulmonary trunk with single branch pulmonary artery (**a**, absent left pulmonary artery; **b**, absent right pulmonary artery).*

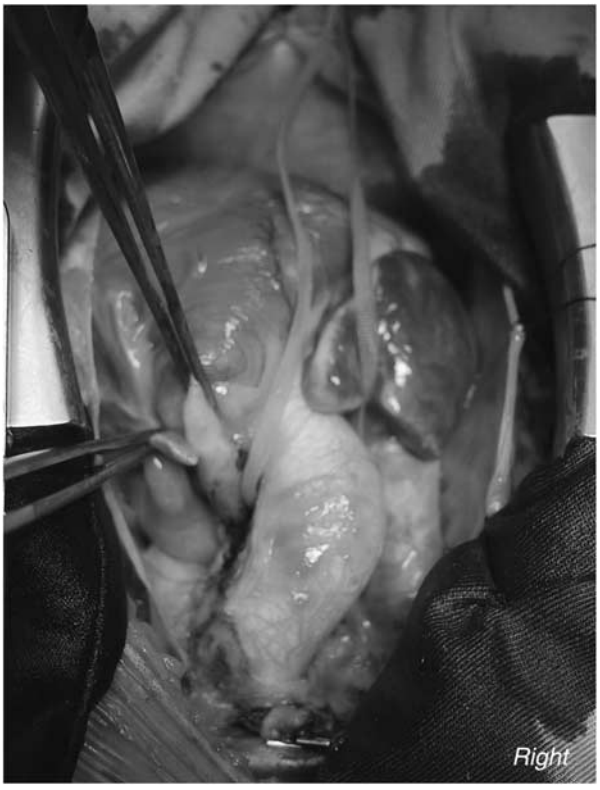

Figure 3.
Peri-operative view showing the main and the right pulmonary artery and the absent left pulmonary artery.

Urgent modified Blalock–Taussig shunt operations were performed in two patients with very low pulmonary indices (Fig 4) and hypoxic spells leading to haemodynamic compromise. The shunts revealed systolic and diastolic gradients of 5- and 7-millimetre mercury gradients, respectively, in the first patient and 4- and 8-millimetre mercury gradients, respectively, in the second patient and provided room-air oxygen saturations of 71% and 79% in the corresponding patients. Both patients were followed up for 2 days at the intensive care unit and discharged home on the seventh post-operative day.

There was one patient – the third patient on the table – who received an elective modified Blalock–Taussig shunt between the left subclavian artery and the left pulmonary artery. The child was extubated at the intensive care unit early post-operatively; however, re-intubation was required the next day owing to pulmonary overflow despite forced diuresis and inotropic support. Further extubation attempts failed and he underwent catheterization 1 week after the operation. The catheterisation showed the patent shunt and relatively enlarged left pulmonary artery (Supplementary materials 2 and 3). We decided to perform a total correction after explaining the risks to the family and following their consent. The atrial septal defect was left open. A small hole was created on the ventricular septal defect Dacron patch with a 4-millimetre punch and a check-valve system was created. The right ventricular outflow tract was reconstructed with an autologous transannular pericardial patch extending to the left pulmonary artery to enlarge the vessel. Weaning off the cardiopulmonary bypass could not be possible after three attempts due to suprasystemic right ventricular pressure and extracorporeal membrane oxygenator was instituted. The sternum was not closed. The extracorporeal membrane oxygenator support was discontinued after 5 days and the chest was closed on the seventh day. He was extubated 3 days later and discharged from the intensive care unit on the twelfth day. Follow-up was uneventful with 95% room-air oxygen saturation. He was sent home after 1 week. The intensive care unit stay was 28 days and total hospital stay was 38 days for the particular patient.

The remaining two patients received total correction uneventfully (Fig 5). The duration of cross-clamp and cardiopulmonary bypass times were

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Table 3. Pulmonary artery parameters for surgical decision.

Patient	BSA	Pulmonary annulus (mm)	Main PA diameter (mm)	Branch PA diameter (mm)	McGoon index	Nakata index	Pulmonary valve Z-score	Main PA Z-score	Branch PA Z-score
1	0.32	5	6	9.2	1.3	86.3	−4.86	−1.15	−1.65
2	0.29	6	6	9.4	1.3	103.8	−3.37	−1.19	−1.68
3	0.45	7	8	1st: 11; 2nd: 15	1st: 1.4; 2nd: 1.8	1st: 160.3; 2nd: 192.4	−3.76	−1.21	1st: −1.64; 2nd: −1.63
4	0.68	7.3	9.8	17	2.1	291.5	−5.13	−1.12	−1.72
5	0.95	13	16	18	1.9	240.1	−2.17	−1.38	−1.65

BSA = body surface area; mm = millimetre; PA = pulmonary artery

Table 4. Operative and post-operative data.

Patient	Surgery	t-CC (min.)	t-CPB (min.)	Mechanical ventilation (hrs.)	ICU stay (days)	Hospital stay (days)	sO_2 (%)
1	R m-BT shunt	N/A	17	6.4	2	9	71
2	R m-BT shunt	N/A	22	8.2	2	9	79
3	L m-BT shunt + total correction (check-valved VSD patch)	N/A for the 1st operation; 2nd: 56	1st: 19; 2nd: 180	1st: 4; 2nd: 584	28	38	95
4	Total correction	44	67	5.2	1	7	100
5	Total correction	28	41	4.4	1	7	100

hrs. = hours; ICU = intensive care unit; L = left; m-BT = modified Blalock–Taussig; min. = minutes; R = right; sO2 = oxygen saturation; t-CC = cross-clamp time; t-CPB = cardiopulmonary bypass time; VSD = ventricular septal defect

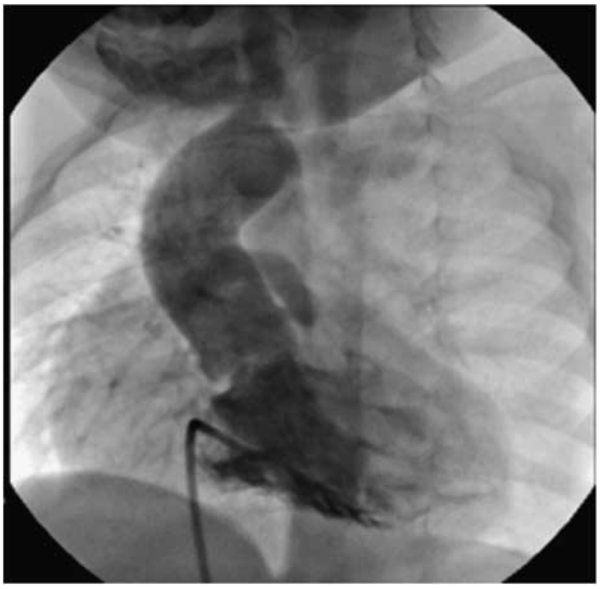

Figure 4.
Angiography showing a small single pulmonary artery unsuitable for total correction in a patient who underwent shunt procedure.

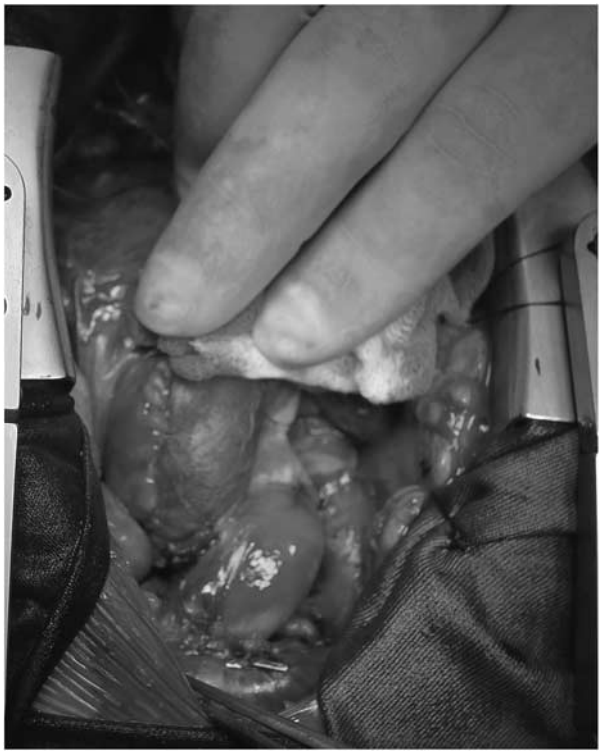

Figure 5.
Peri-operative view following surgical correction of Tetralogy of Fallot and single pulmonary artery.

44 and 67 minutes, respectively, for the fourth patient on the table and 28 and 41 minutes, respectively, for fifth patient on the table. In both patients, weaning off the cardiopulmonary bypass was possible with low-dose inotropic support (5 micrograms per kilogram per minute dopamine). Right ventricle to left ventricle pressure ratios were 0.79 and 0.72 at the early period, correspondingly. There was one patient who stayed 2 days and another who stayed 1 day at the intensive care unit and both were discharged home after 7 days.

Mortality did not occur. Morbidity, with a rate of 20%, including prolonged ventilatory support, consecutive angiography and surgery, and extra-corporeal membrane oxygenator support, occurred in one patient. Mean durations of hospital stay and follow-up were 14 plus or minus 13.4 days and 184.5 plus or minus 89.3 days, respectively. The patients who received palliative procedures are on the list for total correction. The patients who received total correction are followed up regularly at the outpatient clinics symptom free, with normal physical and psychosocial growth and performing normal daily-life activities.

Discussion

Unilateral absent pulmonary artery, as an isolated defect, accounted for less than 1% among congenital malformations.[1,2] It may be associated with other cardiovascular pathologies and the most common lesion encountered in association with unilateral absent pulmonary artery is the Tetralogy of Fallot. Around 1–3% of patients with Tetralogy of Fallot present with single pulmonary artery branch.[4,5] Other congenital malformations that might accompany with unilateral absent pulmonary artery are septal defects, coarctation of the aorta, subvalvular aortic stenosis, transposition of the great arteries, and pulmonary stenosis.[2] The incidence of the absence of the left pulmonary artery is higher,[1,2,4,5] although absent right pulmonary artery has also been reported.[4,6] Our modest series included four cases of absent left pulmonary artery and a single case of right pulmonary artery absence.

Several theories are speculated for the emergence of the pathology; however, yet, none is sufficient to precisely define the pathophysiological mechanisms. For example, during the involution of the right or the left sixth arch corresponding right or the left pulmonary may also disappear.[7] On the other hand, the issue is not sufficient to prove the association of Tetralogy of Fallot with absent left pulmonary artery.[5] Another hypothesis relates to the abnormal division of the truncus. Dorsal shift of the ridges may result in development failure of the associated pulmonary artery. This is the ontogenetic theory and may explain the absence of the right pulmonary artery being frequent as an isolated defect rather

https://doi.org/10.1017/S1047951112000911 Published online by Cambridge University Press

than associated with other malformations, as well as the frequency of the absence of the left pulmonary artery in the Tetralogy of Fallot pathology.[8] However, cases of Tetralogy of Fallot with absent right pulmonary artery are also present and ontogenetic theory fails in such cases.[5] Other than the genetic basis, acquired factors may lead to the loss of one pulmonary artery. Severe cyanosis in the course of Tetralogy of Fallot frequently goes with erythrocytosis and increased viscosity of the blood. Addition of those factors to decreased pulmonary blood flow may cause pulmonary thrombosis[9] and eventual arterial sequestration owing to long-standing thrombus. If the phenomenon is the case, at least remnants of the pulmonary vasculature should be present. None of our patients had pulmonary artery loss secondary to occlusion and all had true absence.

Patients with unilateral absent pulmonary artery may present with a wide range of symptoms. Although rarely, they can be asymptomatic or present to the clinic with symptoms related to severe pulmonary hypertension and congestive cardiac failure including compromised functional status, dyspnoea, and even haemoptysis.[2,3] The severity of the symptoms is also related to the accompanying cardiac malformation. In case of Tetralogy of Fallot, major symptoms are cyanosis and cyanosis-related further complications.

The reported number of patients with Tetralogy of Fallot and single pulmonary artery is not high. The pathology may easily be suspected during echocardiography and clearly identified with conventional angiography, as well as magnetic resonance or computerised tomography measures. Pulmonary venous wedge angiography is useful to investigate the presence of a hilar pulmonary artery and intrapulmonary vessels.[3] All are also valuable for surgical strategy as well.

Surgery options for Tetralogy of Fallot with absent pulmonary artery include either conventional staged approach or single-stage total correction.[10] Both options are not event free and with considerable mortality and morbidity rates. In a recent study by Bockeria et al,[5] 5% mortality rate after palliative and corrective procedures, individually, were reported. Zhang et al[4] indicated higher rates such as 18.75% mortality (two early, one late death) in their cohort after total correction. In the updated world literature in 2007, among 100 patients with Tetralogy of Fallot having unilateral absent pulmonary artery 20% received palliative procedures and the mortality rate reached up to 35%, whereas patients receiving total correction was less than 70% with a mortality rate of approximately 8%.[5] On the other hand, the patients with severe pulmonary hypertension and congestive cardiac failure are usually late cases and do not benefit from corrective surgery. In such cases, palliative methods with vasodilators and embolisation or even pneumonectomy in patients with debilitating haemoptysis may be effective.[3] Heart and lung transplantation can be another alternative.

Our cohort included five patients with diverse symptoms, body features, and pulmonary indices (Table 1). For the first two cases, our decision was a palliative procedure considering their emergency status, weights, body mass indices, and pulmonary sizes (Tables 1, 3, 4). Although the last two patients had unilateral absent pulmonary artery, their ages, weights, body mass indices, and pulmonary artery sizes were suitable for total correction; hence, these cases received single-stage uneventful corrective surgery (Tables 1, 3, 4). The third patient was the most challenging case in the group. His demographic features were suitable for total correction (Table 1); however, pulmonary indices indicated a very high risk for total correction (Table 3). Thus, we preferred a left-sided modified Blalock–Taussig shunt (Table 4). Although weaning from mechanical ventilator could be possible, re-entubation necessitated owing to the overflow phenomenon. Consecutive angiography showed increased pulmonary artery size (Table 3). Despite the risks, we performed total correction with fenestrated Dacron patch closure of the ventricular septal defect. Survival could be possible with extracorporeal membrane oxygenator support and careful intensive care management. He has been well and followed up, with New York Heart Association Class I–II symptoms after hospital discharge.

In conclusion, Tetralogy of Fallot with unilateral absent pulmonary artery is a rare variant. Our modest series indicated the feasibility of surgical correction in patients with appropriate unilateral pulmonary artery size and palliative procedures when the pulmonary artery size is smaller than that predicted for the age and body surface area. Both palliative and corrective procedures are not event free and include certain morbidity rates. Long-term follow-up results of larger series are warranted in order to achieve an established treatment protocol.

Acknowledgement

The authors would like to thank Mr Robert Moore for the linguistic revision of the manuscript.

Supplementary materials

For supplementary materials referred to in this article, please visit http://dx.doi.org/doi:10.1017/S1047951112000911

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