Anomalous branch of pulmonary artery from the aorta and tetralogy of Fallot: morphology, surgical techniques and results

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Abstract

OBJECTIVES: Tetralogy of Fallot (TOF) with hemitruncus (HT) is a rare entity. In this report, we present our experience with this condition over the last 20 years.

METHODS: Between January 1994 and June 2013, 11 patients with HT and TOF underwent surgery at the All India Institute of Medical Sciences, New Delhi, India. All available clinical, radiographic, echocardiographic, cardiac catheterization, operative and follow-up data were reviewed.

RESULTS: The mean age was 73 ± 7.1 months (range 7 months to 18 years) and the mean weight was 15.7 ± 1.2 kg. The mean preoperative saturation was 79.3 ± 11.7% (range 62–92%). Six patients had anomalous left pulmonary artery (PA), whereas 5 had an anomalous right PA arising from the aorta. Surgical procedures consisted of complete intracardiac repair of TOF with direct implantation of the anomalous PA into the main PA (n = 7), intracardiac repair of TOF with an interposition saphenous vein graft between the right PA and main PA (n = 1), and reconstruction of the left PA with autologous pericardium with intracardiac repair of TOF (n = 1), direct implantation of the anomalous PA into the main PA with an innominate to right pulmonary artery shunt (n = 1) and a right PA banding with innominate to left PA shunt (n = 1). There were two early deaths. Follow-up ranged from 3 to 73 months. All survivors are in NYHA Class I and follow-up echocardiograms did not show any residual lesions.

CONCLUSIONS: Surgical repair of HT with TOF results in acceptable early outcomes. The surgical strategy needs to be individualized to the anatomy of the patient.

Keywords: Tetralogy of Fallot • Hemitruncus • Anomalous pulmonary artery

INTRODUCTION

The association of anomalous origin of a branch of pulmonary artery (PA) from the aorta (AOPA), often referred to as hemitruncus (HT), and tetralogy of Fallot (TOF) is rare [1–5]. The anomaly has been described in the form of isolated case reports, but larger series of the incidence, natural history, haemodynamics and morphology are not available. The anomaly has been described to be successfully repaired by many in the larger series of patients with TOF [1], but no specific management guidelines exist. In this report, we describe our experience with this condition over the last 20 years.

PATIENTS AND METHODS

Between January 1994 and June 2013, a total of 5241 patients with TOF underwent intracardiac repair at the All India Institute of Medical Sciences, New Delhi, India. Of these, 11 patients underwent surgery for HT and TOF (Table 1), an incidence rate of 0.21%. The study protocol was duly approved by the institute ethics committee. All available clinical, radiographic, echocardiographic and cardiac catheterization data were reviewed from the available medical records. Operation records were analysed in detail and the morphological findings studied. The intensive care unit (ICU) charts were reviewed to study the postoperative course. Details of hospital survival, ventilator and inotropic support and duration of hospital stay were recorded. The follow-up protocol included outpatient visits at 1 week, 1 month, 3 months, 6 months and then yearly intervals. Specific information sought at follow-up visits included adequacy of surgical repair, details of ventricular function and any residual gradients/regurgitation. Data are presented as simple numbers with mean and median values as applicable.

RESULTS

Of the 11 patients, 8 (72.8%) were male. The mean age was 73 ± 7.1 months (range 7 months to 18 years) and the mean
weight was 15.7 ± 1.2 kg. All patients except patient 8 presented with cyanosis. Except patients 2 and 11, cyanosis manifested after the age of 1 year in these patients. Patients 2 and 11 had a history of recurrent cyanotic spells. No patient had congestive cardiac failure. Chest radiographs in all patients showed differential vascularity and plethora on the side with anomalous connection of PA. The mean preoperative saturation was 79.3 ± 11.7% (range 62–92%). Transthoracic echocardiography was performed in all patients and demonstrated findings typical of TOF. In 9 (81%) patients, it could clearly demonstrate a diagnosis of associated AOPA, whereas in 2 patients it raised the suspicion of AOPA. Cardiac catheterization and cine angiography was performed in all patients for anatomical and haemodynamic assessment under conscious sedation (Table 2 and Figs 1–4). Expectedly, the right ventricular systolic pressure was equal to the left ventricular systolic pressure owing to the presence of a large ventricular septal defect in all patients. Also, consistent with the diagnosis of TOF, all patients had right ventricular outflow tract obstruction. Reliable estimation of pulmonary obstruction with the PA connected to the right ventricle was available in 8 of 11, i.e. 72% patients. On the other hand, in only 4 (36%) patients: nos. 6, 7, 8 and 9, was it possible to enter the anomalously connected PA for haemodynamic assessment. The pulmonary vascular resistance index (PVRI) in the lung with anomalous origin of the PA was extremely high except in patient 10. There was stenosis at the origin of the anomalously connected right PA in patient 10 and consequently the PVRI of the right lung was calculated to be 7.7 WU m². In all patients, right ventricular angiogram and/or aortogram confirmed the anomalous origin of the PA from the ascending aorta. Occasionally, pulmonary arteries appeared confluent on initial right ventricular angiogram as in patient 7 (Fig. 2A). Nevertheless, contrast injection made in the ascending aorta demonstrated the anomalous origin of the PA from the ascending aorta (Fig. 2B). Computed tomography angiography was performed additionally in 4 patients (5, 7, 8 and 9) for better delineation of the anatomy prior to surgical repair. Patient 10 was diagnosed to have TOF with right pulmonary artery (RPA) atresia on preoperative investigations; however, at operation, there was a small stump of the RPA arising from the ascending aorta with a 2-cm atretic segment followed by re-formation of a 4-mm RPA at the right lung hilum.

Morphology

Six patients had an anomalous left pulmonary artery (LPA), whereas 5 had an anomalous RPA arising from the aorta. In 7 patients, the anomalous PA took its origin from the left posterolateral aspect of the ascending aorta. In 4 patients, it originated from the rightward and posterolateral aspect of the ascending aorta, just proximal to the take-off of the innominate artery. The peculiar morphological findings in patient 10 are described above. The origin and proximal course of coronary arteries were normal in all patients except patients 5 and 8. Patient 5 had a single coronary

### Table 1: Summary of patients with tetralogy of Fallot and hemitruncus

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (months)</th>
<th>Weight (kg)</th>
<th>Preop saturation (%)</th>
<th>Anomalous artery</th>
<th>Site of origin</th>
<th>Arch</th>
<th>Surgical procedure</th>
<th>ICU stay (days)</th>
<th>Outcome</th>
<th>Follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>18/M</td>
<td>7</td>
<td>89</td>
<td>RPA</td>
<td>RPL</td>
<td>Left</td>
<td>HR + TC</td>
<td>2</td>
<td>Survived</td>
<td>49</td>
</tr>
<tr>
<td>2</td>
<td>7/M</td>
<td>4.5</td>
<td>62</td>
<td>LPA</td>
<td>LPL</td>
<td>Left</td>
<td>HR + TC</td>
<td>1</td>
<td>Died</td>
<td>Died</td>
</tr>
<tr>
<td>3</td>
<td>72/F</td>
<td>19.5</td>
<td>84</td>
<td>LPA</td>
<td>LPL</td>
<td>Left</td>
<td>HR + TC</td>
<td>2</td>
<td>Survived</td>
<td>18</td>
</tr>
<tr>
<td>4</td>
<td>156/F</td>
<td>40</td>
<td>91</td>
<td>RPA</td>
<td>RPL</td>
<td>Right</td>
<td>HR + TC</td>
<td>2</td>
<td>Survived</td>
<td>56</td>
</tr>
<tr>
<td>5</td>
<td>156/M</td>
<td>20</td>
<td>90</td>
<td>RPA</td>
<td>RPL</td>
<td>Right</td>
<td>HR + TC</td>
<td>2</td>
<td>Survived</td>
<td>73</td>
</tr>
<tr>
<td>6</td>
<td>60/M</td>
<td>12</td>
<td>85</td>
<td>RPA</td>
<td>RPL</td>
<td>Right</td>
<td>PA band + innominate to LPA shunt</td>
<td>2</td>
<td>Survived</td>
<td>5</td>
</tr>
<tr>
<td>7</td>
<td>42/M</td>
<td>10</td>
<td>72</td>
<td>LPA</td>
<td>LPL</td>
<td>Right</td>
<td>HR + TC (LPA augmented with pericardium)</td>
<td>2</td>
<td>Survived</td>
<td>3</td>
</tr>
<tr>
<td>8</td>
<td>216/M</td>
<td>40</td>
<td>92</td>
<td>RPA</td>
<td>RPL</td>
<td>Left</td>
<td>HR + TC</td>
<td>2</td>
<td>Survived</td>
<td>3</td>
</tr>
<tr>
<td>9</td>
<td>33/F</td>
<td>10.3</td>
<td>78</td>
<td>LPA</td>
<td>LPL</td>
<td>Left</td>
<td>HR + TC</td>
<td>2</td>
<td>Survived</td>
<td>6</td>
</tr>
<tr>
<td>10</td>
<td>36/M</td>
<td>12</td>
<td>66</td>
<td>RPA</td>
<td>RPL</td>
<td>Left</td>
<td>HR + TC (saphenous vein graft)</td>
<td>2</td>
<td>Survived</td>
<td>17</td>
</tr>
<tr>
<td>11</td>
<td>7/M</td>
<td>5.8</td>
<td>63</td>
<td>LPA</td>
<td>LPL</td>
<td>Left</td>
<td>HR + innominate to RPA shunt without bypass</td>
<td>6 h</td>
<td>Died</td>
<td>Died</td>
</tr>
</tbody>
</table>

FA: femoral artery; LPA: left pulmonary artery; HR: hemitruncus repair; TC: total correction; TOF: tetralogy of Fallot.

### Table 2: Cardiac catheterization data of patients with tetralogy of Fallot and hemitruncus

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (months)</th>
<th>Weight (kg)</th>
<th>Preop saturation (%)</th>
<th>Anomalous artery</th>
<th>Pressure in normally connected PA (sys/dias/mean)</th>
<th>PVRI (WU m²) of lung with anomalous origin of PA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>18/M</td>
<td>7</td>
<td>89</td>
<td>RPA</td>
<td>Not entered</td>
<td>Not calculated</td>
</tr>
<tr>
<td>2</td>
<td>7/M</td>
<td>4.5</td>
<td>62</td>
<td>LPA</td>
<td>Not entered</td>
<td>Not calculated</td>
</tr>
<tr>
<td>3</td>
<td>72/F</td>
<td>19.5</td>
<td>84</td>
<td>LPA</td>
<td>44/22 (mean 30)</td>
<td>Not calculated</td>
</tr>
<tr>
<td>4</td>
<td>156/F</td>
<td>40</td>
<td>91</td>
<td>RPA</td>
<td>30/17 (mean 22)</td>
<td>Not calculated</td>
</tr>
<tr>
<td>5</td>
<td>156/M</td>
<td>20</td>
<td>90</td>
<td>RPA</td>
<td>36/20 (mean 27)</td>
<td>Not calculated</td>
</tr>
<tr>
<td>6</td>
<td>60/M</td>
<td>12</td>
<td>85</td>
<td>RPA</td>
<td>22/13 (mean 16)</td>
<td>42.7</td>
</tr>
<tr>
<td>7</td>
<td>42/M</td>
<td>10</td>
<td>72</td>
<td>LPA</td>
<td>29/16 (mean 20)</td>
<td>37.8</td>
</tr>
<tr>
<td>8</td>
<td>216/M</td>
<td>40</td>
<td>92</td>
<td>RPA</td>
<td>36/18 (mean 26)</td>
<td>7.7</td>
</tr>
<tr>
<td>9</td>
<td>33/F</td>
<td>10.3</td>
<td>78</td>
<td>LPA</td>
<td>24/14 (mean 19)</td>
<td>Not calculated</td>
</tr>
<tr>
<td>10</td>
<td>36/M</td>
<td>12</td>
<td>66</td>
<td>RPA</td>
<td>Not entered</td>
<td>Not calculated</td>
</tr>
<tr>
<td>11</td>
<td>7/M</td>
<td>5.8</td>
<td>63</td>
<td>LPA</td>
<td>Not entered</td>
<td>Not calculated</td>
</tr>
</tbody>
</table>

FA: femoral artery; PA: pulmonary artery; PVRI: pulmonary vascular resistance index.
artery arising from the right coronary sinus, while patient 8 had a left anterior descending artery arising from the right coronary sinus before crossing the right ventricular outflow tract.

Surgical procedures

In all patients, surgical procedures were performed via a standard median sternotomy. Surgical procedures included intracardiac repair of TOF and direct implantation of the anomalous PA to the main PA (MPA) \((n = 7)\), intracardiac repair of TOF and interposition saphenous vein graft to join the right PA to the MPA \((n = 1, \text{ Patient 10})\), and reconstruction of the LPA with autologous pericardium with intracardiac repair of TOF \((n = 1, \text{ Patient 7})\). One patient (no. 11) underwent direct implantation of the anomalous PA into the MPA with an innominate to RPA shunt \((n = 1, \text{ Patient 11})\), while in patient 6 RPA band with an innominate to LPA shunt was performed. Intracardiac repair in all patients was performed with the use of cardiopulmonary bypass (CPB), while patients 6 and 11 underwent the surgical procedure without CPB. Standard hypothermic CPB with aorto-bicaval cannulation at 28°C was used. Prior to establishing CPB, the MPA, branch PAs and the anomalous

Figure 1: Cine angiogram of patient 6 shows the right pulmonary artery (PA) arising from the left posterolateral aspect of the ascending aorta. The main PA continues as a small-sized left PA.

Figure 2: Cine angiogram of patient 7. (A) On right ventricular injection in the anteroposterior view, both the pulmonary arteries appear to be confluent. (B) An aortic root injection in the lateral view clearly demonstrates the left pulmonary artery coming off the ascending aorta.
PA were looped. Upon establishing CPB, the snare around the anomalous PA was tightened to prevent run-off into the pulmonary circulation; the patent ductus arteriosus/ligamentum arteriosum was divided.

Before achieving cardioplegic arrest, the aim was to disconnect the anomalous PA from the aorta and to achieve continuity between both the LPA and the RPA. Wherever possible, a direct anastomosis between the anomalous PA and the MPA was a preferred option. However, in cases where there was an intervening stenotic/atretic segment between the aorta and the anomalous PA that would prevent adequate mobilization and a tension-free anastomosis with the contralateral PA, an interposition graft was preferred. To achieve this, the anomalous PA was clamped and divided at its aortic origin. The aortic end was repaired. The divided PA was passed underneath the aorta for a more physiological lie. The MPA was opened after applying side-biting clamps and an end-to-side anastomosis was fashioned between the divided PA and the MPA. Following this, the aortic cross-clamp was applied and after delivering cardioplegia, repair of TOF was accomplished using standard techniques. Of the 9 patients who underwent complete primary repair, one patient (no. 8) had a transatrial repair of TOF with repair of HT. In the remaining 8 patients, a transannular patch was required. Of these, patient 10 underwent primary repair along with interposition of a saphenous vein homograft to the RPA; a transannular patch was placed using a pulmonary homograft bearing a monocusp. In the remaining patients undergoing primary repair, a monocusp valve fashioned out of autologous pericardium was used to prevent free pulmonary regurgitation. The mean CPB time was 127 ± 34 (range 75–175) min, and the mean aortic cross-clamping time was 75.8 ± 18 (range 54–95) min in those patients who underwent complete repair on CPB. In the 2 patients who did not undergo complete repair, the reasons were: (i) small LPA, ≏4 mm in patient 6. In this patient, an innominate to MPA shunt was performed using a 5-mm graft. The presence of a right aortic arch facilitated placement of this shunt on the left side from the base of the innominate artery to the MPA without making an anastomosis on the small LPA. (ii) In patient 11, HT repair along with a 4-mm innominate to RPA shunt was preferred over complete primary repair because of a hypoplastic RPA and poorly developed peripheral pulmonary vasculature.

Figure 3: Cine angiogram of patient 8. (A) The main pulmonary artery (PA) continues as the left PA. (B) An aortic root injection in the lateral view demonstrates the right PA coming off the ascending aorta.

Figure 4: Cine angiograms of patient 11 showing (A) left pulmonary artery arising from the aorta and (B) a hypoplastic right pulmonary artery arising from the right ventricle. Reproduced with permission from Garg et al. [6].
Postoperative course and follow-up

There were two early deaths (Patients 2 and 11). Patient 2 died of severe low cardiac output syndrome on the first postoperative day despite documented adequate surgical repair. Patient 11 developed persistent hypoxia and hypercarbia despite a patent shunt. He had unexplained bradycardia and cardiac arrest 6 h following surgery and could not be resuscitated. In both patients, autopsy could not be performed because the parents did not give their consent.

In the remaining patients, the postoperative course was uneventful. They received elective inotropic support in the form of dopamine 5 µg/kg/min and sodium nitroprusside 0.5 µg/kg/min. The median duration of mechanical ventilatory support was 14 h, and the median ICU stay was 48 h in these patients. The median hospital stay was 8 (range 4–10) days.

At discharge, patient 6 had systemic saturation of 77%. The systemic saturation of the remaining patients was 95–100%. Predischarge echocardiograms revealed normal biventricular function with no gradient at the site of the anastomosis of the anomalous PA to the MPA. There was moderate pulmonary regurgitation in 3 patients and mild PR in 1. The patient who had an RPA band demonstrated a band gradient of 75 mmHg at the site of the band. Follow-up ranged from 3 to 73 (mean 25.6 ± 11.7) months and was 100% complete. All survivors are in NYHA Class I and follow-up echocardiograms did not show any gradients at the anastomosis sites. Pulmonary regurgitation and RV dimensions have not progressed. Patient 8 had a follow-up MRI (Fig. 5) that demonstrated satisfactory repair. None of the patients required reintervention so far. The patient (no. 6) who underwent RPA band with a shunt to the MPA is awaiting cardiac catheterization in preparation for complete repair.

DISCUSSION

The association of HT and TOF is extremely rare. In a recent retrospective single-centre study of 2235 patients with TOF, 8 (0.4%) had HT [1]. Other than this report, TOF with HT in the literature is limited to case reports [2–10]. Nonetheless, surgical technique and outcome have not been dealt with in detail so far.

When AOPA occurs in isolation, an anomalous PA on the right side is 4–8 times more common than on the left side [8]. Although an anomalous PA can be present on either side in patients with TOF, it has been more commonly seen on the left side [5, 8]. The embryological basis for this anomaly is probably the failure of development of a left sixth arch and persistence of a left fifth arch [3]. Some believe that the anomaly is a result of the defective septation of the primitive truncus arteriosus. Unequal division of the latter probably accounts for a higher incidence of anomalous LPA rather than RPA in patients with TOF [10]. HT should be suspected clinically in any patient with TOF with chest radiograph showing differential vascularity of lung fields owing to differences in the blood flow. Although echocardiography can reliably detect the anomaly, cardiac catheterization and cine angiograms are confirmatory [5]. In patients with HT, while assessing pulmonary vascular resistance, it is important to calculate pressure and blood flow in each lung separately. However, this may not be feasible with accuracy during cardiac catheterization. Therefore, owing to various assumptions made during such calculations, estimation of the PVRI may be fallacious especially in the lung with anomalous origin of the PA from the aorta. Cardiac MRI can provide a reasonable estimate of pulmonary blood flow and the PVRI in the individual lung instead.

Irrespective of the mode of assessment, being exposed to aortic pressure, the PVRI on the side with AOPA is expected to be prohibitively high. Nonetheless, in the setting of TOF, the lung with normally connected PA has a low PA pressure and, consequently, relatively low PVRI. Further, as in hearts with disconnected pulmonary arteries, the resistances are calculated as 1/Total PVR = 1/PVR right + 1/PVR left [10, 11]. Total pulmonary vascular resistance in patients with TOF and HT is likely to be acceptable for surgical repair, provided two lung repair can be achieved surgically. This is further supported by successful surgical repair and good functional recovery in our patients.

Early detection and surgery for these patients is mandatory to prevent development of pulmonary vascular occlusive disease [7]. Considering direct aorto-pulmonary communication, these patients are at risk of rapid progression of pulmonary vascular disease in the lung with the PA connected anomalously to the aorta. Survival
beyond adolescence is rare with only few reports in the literature. Till 2013, only 5 cases had been reported in which the patients survived beyond the first decade of life [2]. In our series, however, nearly one-fourth (23%) of patients presented after the first decade of life. This is in keeping with the trend of late presentation in a developing country like India. Further, although all patients underwent the desired surgical repair, their long-term outcome remains to be seen in view of high pulmonary vascular resistance especially in the lung with an anomalously connected PA.

As has been observed previously, if an anomalous origin of the PA from the aorta is not recognized, an attempted repair of TOF can be disastrous and has led in the past to failure to wean off the patient from CPB, with the anomaly being detected only later on at autopsy [9, 10]. The first reported case of successful surgical correction of TOF with HT was by Morgan [9]. Since then, there have been isolated case reports of successful surgical correction of this anomaly.

We were able to achieve a successful primary repair in all but 2 patients for reasons detailed above. With a proper anatomical delineation, early primary repair appears to be the best strategy in these patients. The principles of repair of this anomaly combine the principles of repair of HT and TOF when being dealt with as isolated anomalies, and surgical reports have not addressed the technical aspects of repair of TOF with HT. We believe that some salient points to be kept in mind are the following: (i) It is preferable to complete all dissection prior to establishing CPB. (ii) If intracardiac repair is not being planned, the shunt procedure and the anastomosis of AOPA to the MPA can be accomplished without CPB. (iii) If repair is planned on CPB, the detachment of AOPA and its anastomosis to the MPA can be achieved prior to aortic cross-clamping after establishing CPB to cut down the aortic cross-clamp time. (iv) We have observed that often the pulmonary annulus in these patients is small and the RVOT requires a transannular patch. In the setting of pulmonary hypertension of one lung, and especially if the other PA is smaller, it is best to avoid free pulmonary regurgitation by conservation of the native valve tissue and by use of a monocusp valve to at least prevent free pulmonary regurgitation. (v) Aggressive use of vasodilators is often helpful to achieve a smoother postoperative course in these patients. These drugs help to reduce the PA pressures, the incidence of PA hypertensive crisis, and the force and velocity of the pulmonary regurgitant jet that is detrimental to right ventricular function.

Like any other patient with repaired TOF, these patients need regular follow-up. However, in addition to routine evaluation for pulmonary regurgitation and right ventricular function, the evaluation must include assessment of the anastomosis site of the MPA with the previous anomalous PA.

If possible, differential lung perfusion scans should be performed in follow-up to assess differential lung perfusion. We were unable to perform the same due to financial constraints.

**Study limitations**

This study is a retrospective audit of a limited number of patients who underwent repair for TOF and HT at our institute. The patient population is much older than normally encountered worldwide. The duration of follow-up is short, and therefore only early outcomes can be interpreted. Long-term clinical data and follow-up haemodynamic assessment are lacking.

**CONCLUSION**

Surgical repair of HT with TOF results in acceptable early outcomes. The surgical strategy needs to be individualized to the anatomy of the patient.

**Conflict of interest:** none declared.

**REFERENCES**


