A comprehensive study of congenital unilateral absence of branch pulmonary artery associated with other congenital heart defects and ipsilateral non-unifocalizable major aorto-pulmonary collateral arteries: A single-center retrospective study

Harpanahalli Ravi Ramamurthy¹, Varsha Walavalkar², Satheesh Siddaiah², Sunita Maheshwari³
¹Department of Pediatric Cardiology & Grown Up Congenital Heart Diseases, Army Hospital (Research & Referral), New Delhi, India, ²Department of Pediatric and Grown-Up Congenital Heart Diseases, Narayana Institute of Cardiac Sciences, Bengaluru, Karnataka, India, ³RXDX Healthcare and Teleradiology Solutions, Bengaluru, Karnataka, India

ABSTRACT

Introduction: Congenital unilateral absence of pulmonary artery (UAPA) is a rare congenital anomaly with the complete absence of intrapericardial segment of one of the branch pulmonary arteries. Sixty percent are associated with other congenital heart defects (CHD) that often need correction.

Aim: To analyze the data of patients with UAPA and ipsilateral non-unifocalizable major aortopulmonary collateral arteries (MAPCAs) associated with other CHD to identify the commonly associated CHD, their management strategies and outcomes.

Materials and Methods: Retrospective data of patients admitted for congenital UAPA with other CHD was compiled from hospital records from 2002 to 2015. The associated CHD were categorized as group I with the decreased pulmonary flow and group II with increased pulmonary flow to the unaffected contralateral pulmonary artery. The determinants of their management were analyzed.

Results: Sixty-five patients of UAPA and ipsilateral non-unifocalizable MAPCAs associated with other CHD were identified. Group I had 41 patients and Group II had 24. The most common CHD associated with UAPA was tetralogy of Fallot (TOF) in 31 patients (47.7%). Fifty-three patients underwent surgery, 48 (73.8%) underwent single lung corrective surgery, 5 (7.6%) palliative surgery and 12 (18.4) received no surgery. Four operated patients died in the immediate postoperative period. The lowest Mc Goon ratio and Nakata index to undergo corrective surgery were 1.0 and 87.4 mm²/m². A follow-up of 21 patients was done, among which 11 patients who underwent single-stage corrective surgery, all are in NYHA class II and saturating above 95%.

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INTRODUCTION

Congenital unilateral absence of pulmonary artery (UAPA) is defined as the complete absence of the intrapericardial segment of one of the main branch pulmonary arteries, either left or right.\(^1\) The prevalence of isolated UAPA without associated congenital heart defects (CHD) varies from 1 in 200,000 to 1 in 300,000.\(^2\) The incidence has been reported by another series as 0.6% and 60% of them are associated with other CHD, among which tetralogy of Fallot (TOF) is the commonest.\(^3\) Congenital UAPA with CHDs may be associated with good-sized unifocalizable ipsilateral major aorto-pulmonary collateral arteries (MAPCAs) or small non-unifocalizable MAPCAs. The severity of symptoms depends on the underlying CHD, development of the affected lung, its vascular elements and pulmonary hemodynamics. The evaluation and surgical planning require echocardiography, computed tomography (CT) scan and/or cardiac catheterization for hemodynamic assessment [Figures 1, 2 and Videos 1-3]. An “ideal” correction for UAPA associated with other CHDs consists of restoring adequate blood flow into both lungs from the right ventricle via the main pulmonary artery and complete repair of the intracardiac shunts. The type of repair possible is determined by the anatomy of the MAPCAs supplying the ipsilateral lung, the size of the unaffected contralateral pulmonary artery (CPA) and the pulmonary arterial pressures. The management options range between corrective two lung or single-lung repair, palliative surgery, or medical follow-up.\(^4\) However, the available literature has focused mainly on the management of TOF with UAPA, both with unifocalizable and non-unifocalizable MAPCAs. The prevalence and management of UAPA with CHD other than TOF have been inadequately described. We conducted a retrospective study of patients diagnosed with congenital UAPA and ipsilateral non-unifocalizable MAPCAs associated with other CHD at a high volume pediatric cardiothoracic center in India with respect to the management strategy, the immediate postoperative outcome and limited long-term outcome.

MATERIALS AND METHODS

The retrospective data of patients admitted for evaluation and treatment following diagnosis of congenital UAPA associated with other CHD was retrieved from the hospital medical records between the years 2002 and 2015 ensuring strict patient confidentiality. The study was approved by the institutional ethical committee. The records were retrieved from the hospital database using the search code “absent pulmonary artery” and applying requisite filters. The records of the patients, including the computed tomography (CT) scan, cardiac catheterization, surgical notes and discharge summaries were reviewed. The patients were contacted by the telephone number available in our records. The patients were categorized in two groups; Group I with

Conclusions: Congenital UAPA is a rare anomaly and associated with a variety of CHDs, TOF being the most common. Single lung corrective surgery in patients with ipsilateral non-unifocalizable MAPCAs has good immediate and long term survival.

Keywords: Congenital heart surgery, pulmonary artery, tetralogy of Fallot, truncus arteriosus
the decreased pulmonary flow [TOF physiology, Table 1] and Group II with increased pulmonary flow [Table 1], depending on pulmonary flow to the lung which had its CPA intact and in continuity with the main pulmonary artery. The preoperative pulmonary artery measurements of patients of Group I was compiled from CT scan and cardiac catheterization reports. Their Nakata index and Mc Goon ratio were calculated as follows:

Nakata index = Cross sectional area of CPA (mm²)/body surface area (m²)

Mc Goon ratio = Diameter of CPA (mm)/diameter of descending thoracic aorta at level of diaphragm (mm)

The oximetry data of patients of Group II was analyzed to obtain the mean pulmonary vascular resistance (PVR) indexed to the body surface area and mean of ratio of PVR index (PVRI) and systemic vascular resistance index (SVRI) in the operated and the unoperated patients.

RESULTS

The data of 65 patients with UAPA associated with other CHDs were retrieved. The patients consisted of 38 (58.4%) males and 27 (41.5%) females, male:female ratio of 1.4:1, with a median age of 67 months (interquartile range 19–129 months). Left pulmonary artery was absent

Table 1: Distribution of study population by diagnosis of the underlying congenital heart disease, the treatment (surgical versus non-surgical) advised, distribution of surgical procedures undertaken for patients undergoing surgery and the justification for patients not advised surgery

| The distribution of underlying CHD in the study sample | Distribution of patients based on underlying CHD (n=65), n (%) | Distribution of patients who underwent surgery (n=53), n (%) | Distribution of patients according to surgical procedure (n=53) | Description of surgical procedure undertaken according to the number of patients in previous column | Patients who underwent non-surgical treatment/medical management (n=12), n (%) | Reasons for no surgery being undertaken for the patients on non-surgical treatment |
|-----------------------------------------------------|---------------------------------------------------------------|-------------------------------------------------------------|-------------------------------------------------------------|-------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------|---------------------------------------------------------------------------------|
| TOF                                                | 31 (47.7)                                                     | 29 (54.7)                                                   | 23                                                          | RVOT augmentation with transannular patch with VSD closure*                                                                                     | 1 (8.3)                                                                          | Unfavorable PA anatomy                                                                 |
| TOF with absent pulmonary valves                    | 4 (6.1)                                                       | 4 (7.5)                                                     | 2                                                          | Open pulmonary valvotomy with VSD closure                                                                                                 | 1 (8.3)                                                                          | Right ventricle small and unsuitable for two pump repair                        |
| TOF with absent pulmonary valves                    | 4 (6.1)                                                       | 4 (7.5)                                                     | 1                                                          | Modified BT shunt Rastelli repair to single branch pulmonary artery with VSD closure                                                    | 0                                                                               |                                                                                                |
| DORV with VSD with PS                              | 5 (7.7)                                                       | 3 (5.6)                                                     | 1                                                          | Open pulmonary valvotomy with VSD closure                                                                                                 | 2 (16.6)                                                                         | Non routable VSD, PA pressures unsuitable for single ventricle pathway (patient refused corrective surgery) |
| DORV with VSD with PS                              | 5 (7.7)                                                       | 3 (5.6)                                                     | 1                                                          | Modified BT shunt Glenn to single branch PA                                                                                               | 1                                                                               |                                                                                                |
| CCTGA VSD ASD PS                                   | 1 (1.5)                                                       | 1 (1.8)                                                     | 1                                                          | Glenn to single branch PA                                                                                                                  | 0                                                                               |                                                                                                |
| Group II: Increased pulmonary blood flow to contralateral unaffected lung |                                                                 |                                                             |                                                             |                                                                                                                                             |                                                                                  |                                                                                  |
| Truncus arteriosus                                 | 8 (12.3)                                                      | 4 (7.5)                                                     | 3                                                          | Truncus arteriosus repair with homograft                                                                                                   | 4 (33.3)                                                                         | Severe irreversible PA rendered the patients inoperable                         |
| VSD                                                | 8 (12.3)                                                      | 7 (13.2)                                                    | 7                                                          | VSD closure with patch                                                                                                                    | 1 (8.3)                                                                          | Severe irreversible PA rendered the patient inoperable                         |
| Aorto-pulmonary window                             | 3 (4.6)                                                       | 2 (3.7)                                                     | 2                                                          | Aortopulmonary window repair                                                                                                               | 1 (8.3)                                                                          | Severe irreversible PA rendered the patient inoperable                         |
| ASD                                                | 3 (4.6)                                                       | 2 (3.7)                                                     | 2                                                          | ASD closure                                                                                                                                  | 1 (8.3)                                                                          | Small ASD with asymptomatic patient                                            |
| Cor triatriatum with ASD                           | 1 (1.5)                                                       | 1 (1.8)                                                     | 1                                                          | Excision of cor triatriatum, with ASD closure                                                                                              | 0                                                                               | Moderate PDA with asymptomatic patient                                        |
| PDA                                                | 1 (1.5)                                                       | 0 (0)                                                       | 0                                                          |                                                                                                                                             | 1 (8.3)                                                                          |                                                                                  |

*This one patient had undergone single pulmonary artery banding as first stage surgery at age of one month. ASD: Atrial septal defect, BT shunt: Blalock taussig shunt, CHD: Congenital heart disease, CCTGA: Corrected transposition of great arteries, DORV: Double outlet right ventricle, RVOT: Right ventricular outflow tract, PAH: Pulmonary arterial hypertension, PS: Pulmonary stenosis, PA: Pulmonary artery, TGA: Transposition of great arteries, TOF: Tetralogy of Fallot, VSD: Ventricular septal defect, PDA: Patent ductus arteriosus
in 47 patients (72.3%), 18 (27.6%) had absent right pulmonary artery. One patient of truncus arteriosus with UAPA had partially anomalous pulmonary venous connection and double aortic arch. All our patients had small MAPCAs that were unsuitable for unifocalization supplying the ipsilateral lung. Hypoplasia of the affected lung was seen on radiographs and thoracic CT scan in 15 patients (23%). The epidemiological parameters and the common cardiac abnormalities in addition to the underlying CHD are enumerated in Table 2.

The most common underlying CHD associated with congenital UAPA and non-unifocalizable MAPCAs was TOF in 31 (47.7%) patients. The other CHD’s associated are enumerated in Table 1.

Group I with decreased pulmonary flow had 41 patients, 33 were offered corrective surgery and three unidirectional Glenn shunt, out of which one patient each opted for medical management. Three patients underwent modified Blalock–Taussig (BT) shunt. Two patients were offered no surgery. The accurate measurements of the pulmonary artery were retrieved in 24 patients. The mean Mc Goon ratio and mean Nakata index of CPA among these patients was 1.3 ± 0.55 and 271.8 ± 296.4 mm²/m². The lowest Mc Goon ratio that successfully underwent single lung corrective surgery was 1.0 and Nakata index of 87.4 mm²/m².

Group II, with increased pulmonary flow, had 24 patients and the oximetry data of 20 was retrieved. 16 patients received corrective surgery. The oximetry data analysis of 12 of these patients revealed a mean PVRI of 3.5 ± 2.7 wood units and mean PVRI: SVRI ratio of 0.18 ± 0.18. Four patients were deemed operable on echocardiography and cardiac catheterization was not carried out. Eight patients were offered medical management because of irreversible pulmonary artery hypertension with mean PVRI of 11.3 (±9) wood units and mean PVRI: SVRI ratio of 0.44 (±0.2) [Table 1 and Figure 3].

The management approach to the patients of both groups is depicted in Figure 3. The distribution of the underlying CHD in all 65 patients, their management decision taken, i.e., surgical versus medical, the surgical procedures undertaken in 53 patients and reasons for medical management in 12 patients are tabulated in Table 1. Four patients died in the immediate postoperative period. Twenty one patients could be followed up by their review records and by telephonic interview. The mean age of these 21 patients was 87.6 months (±75) at first diagnosis. Five received no surgery nor intervention. After 10 years, four are in NYHA class II. The oldest surviving patient is a case of truncus arteriosus with UAPA aged 25 years. One patient aged 18 years with Eisenmengerized PDA to the CPA is in NYHA class 4 and awaiting heart-lung transplant. Among the 16 followed up patients who underwent surgery, four had BT shunt and one of them could be offered corrective surgery after 3 years. One patient with truncus arteriosus underwent pulmonary artery banding at 1 month of age followed by corrective single lung Rastelli surgery after 3 years and is in NYHA class I and saturating 95% after 10 years. The average interval between the 1st and 2nd stage surgery was 54 ± 65 months. The 11 patients who underwent single-stage corrective surgery, all are in NYHA class I and saturating above 95%. The follow-up data are summarized in Figure 4. None of the patients who could be followed up underwent a repeat cardiac catheterization at our center.

Table 2: The distribution of study population with respect to the epidemiological parameters and common cardiac associations in addition to underlying congenital heart disease

| Distribution parameter                        | Total numbers (n=65), n (%) | Number of patients who underwent surgical treatment (n=53), n (%) | Number of patients who underwent nonsurgical treatment (n=12), n (%) |
|---------------------------------------------|---------------------------|-----------------------------------------------------------------|-----------------------------------------------------------------|
| Distribution of patients by gender          |                           |                                                                  |                                                                 |
| Male                                        | 40 (61.5)                 | 33 (62.2)                                                       | 7 (58)                                                         |
| Female                                      | 25 (38.4)                 | 20 (37.7)                                                       | 5 (42)                                                         |
| Distribution of patients by age (in months) | 84.2±80.5                 | 75.1±81.7                                                       | 124.3±63                                                       |
| Distribution of patients by cardiac position|                           |                                                                  |                                                                 |
| Dextrocardia                                | 60 (92.3)                 | 49 (92.4)                                                       | 11 (91.6)                                                      |
| Levocardia                                  | 4 (6.1)                   | 4 (7.5)                                                         | 0                                                              |
| Mesocardia                                  | 1 (1.5)                   | 0                                                               | 1 (8.3)                                                        |
| Distribution of patients by visceral situs   |                           |                                                                  |                                                                 |
| Situs solitus                               | 63 (96.9)                 | 51 (96.2)                                                       | 12 (100)                                                       |
| Situs inversus                              | 2 (3)                     | 2 (3.7)                                                         | 0                                                              |
| Distribution of patients by atrioventricular concordance | |                                                                  |                                                                 |
| Atrioventricular concordance                | 63 (96.9)                 | 51 (96.2)                                                       | 12 (100)                                                       |
| Atrioventricular discordance                | 2 (3)                     | 2 (3.7)                                                         | 0                                                              |
| Distribution of patients by anatomy of aortic arch | |                                                                  |                                                                 |
| Right arch                                  | 24 (36.9)                 | 20 (37.7)                                                       | 4 (28.5)                                                       |
| Left arch                                   | 40 (61.5)                 | 32 (60.3)                                                       | 8 (71.4)                                                       |
| Double aortic arch                          | 1 (1.5)                   | 1 (1.8)                                                         | 0                                                              |

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DISCUSSION

The paucity of data in congenital UAPA associated with other CHD is explained by its rarity and heterogeneity. We noted that UAPA was associated with a variety of CHD at our center and among our patients, none had MAPCAs that were suitable for unifocalization. Our study therefore has the data of only those patients with UAPA associated with CHD with non-unifocalizable MAPCAs. This may be because our center is a referral center for pediatric cardiac surgery and the patients being operated on here were only the complex ones leading to a selection bias. There were also no data of those patients with isolated UAPA without associated CHD from our center, perhaps because our center would not admit any patients who were not considered for surgical intervention. Thus, 65 patients with UAPA with non-unifocalizable MAPCAs associated with other CHD admitted for management at our center over 13 years were included. This includes only inpatient data and does not reflect the community prevalence of isolated UAPA nor UAPA with CHD. This anomaly has received attention during the last 15–20 years owing to better diagnosis and advances in surgical techniques. Bockeria et al. reported that 67% of patients with UAPA had associated CHD. However, it has been poorly studied in India, despite having a sizeable population with CHD. Trivedi KR et al. reported a series of 45 patients of UAPA over 30 years. They noted that 29 (44%) were associated with other CHD among which TOF was the most common in 12 (41.3%). We noted a similar distribution with 31 (47.7%) patients having TOF and 41 (63%) with TOF physiology in our study [Table 1]. It was interesting to note that the second most common CHD associated with UAPA was truncus arteriosus in eight patients (12.3%) [Table 1]. The association of UAPA with non-unifocalizable MAPCAs and CHD with increased pulmonary flow has not been discussed earlier. Irrespective of the associated CHD, all our patients underwent systematic evaluation as described [Figure 3] with an intent to provide the best possible treatment. It is evident that the first step in the management algorithm was identifying them as low pulmonary flow vs a high pulmonary flow physiology; therefore we categorized our cohort into Group I and group II for this study [Table 1 and Figure 3]. In patients with TOF physiology, as reported by Bockeria et al. as well as Ugurlucan M et al., the pulmonary artery anatomy of the CPA is an important deciding factor for corrective surgery. We too noted the same in the data of 24 patients we could retrieve in Group I where the mean Mc Goon ratio was 1.3 ± 0.55. The lowest Mc Goon ratio and Nakata index that underwent successful single-stage correction were 1.0 and 87 mm²/m², respectively. In the series of 27 patients of Brockeria et al., the group of patients with Nakata index Z score between normal to –6 corresponding to mean Mc

Figure 3: The flowchart depicting the approach to the management of patients with unilateral absence of pulmonary artery with congenital heart diseases in our study
Goon ratio between 1.79 ± 0.44-0.99 ± 0.07 underwent corrective surgery. The number of patients from Brockeria et al. and our series, although limited, provide valuable information as to the possible management decisions based on the size of CPA for this rare anomaly. However, drawing a statistically significant conclusion is difficult. Prospective data are wanting in this subset of patients to accurately determine the cut-off values of Mc Goon ratio and Nakata index. It is imperative to note here that several other factors would have also had a bearing on the decisions taken but beyond the scope of this retrospective study. Some workers have highlighted the importance of left ventricular adequacy before complete correction.\[4\] In Group II, the PVR and the ratio of pulmonary to systemic vascular resistance was taken for deciding the operability as for patients with normal PAs [Table 1].\[8\] There are no established separate PVR values to decide for operability in patients with UAPA with the increased pulmonary flow. There is a possibility that the ipsilateral lung would be contributing to the PAH postoperatively, the extent of which cannot be estimated. Therefore, in our study, patients with a safe margin of PVRI: SVRI ratio of 0.18 ± 0.18 was selected for corrective surgery.

The management based on this approach at our center could offer corrective single lung repair to 53 (81.3%) patients with immediate postoperative mortality of four (7.5%), which is comparable to other studies [Table 3].\[5,9,10\] A series by Zhang et al. from China reported 24 patients operated for TOF with UAPA over 30 years with immediate mortality in two patients [Table 3].\[10\] Till date, the reported largest experience in the management of UAPA associated with TOF is from Bockeria et al., where 27 patients were operated, of which seven (25%) underwent palliation and 20 (75%) underwent complete two lung repair.\[5\] In another series from the same center, 29 patients underwent palliative surgery of which 17 (59%) could undergo complete two lung repair at a later stage.\[9\] Our center being a tertiary care referral hospital, has patients referred from all over India and abroad, thereby providing a large cohort of patients with this rare anomaly. However, the disadvantage of the same was noted during follow-up. Almost 66% of patients were lost to follow up. Among the 21 patients that could be followed up for almost 10 years, all patients who received single lung corrective repair were living a productive life and in NYHA class I [Figure 4]. This is comparable to the

| Authors          | Number of patients | Number of patients with complete repair | Number of patients with palliation only | Early mortality (%) |
|------------------|--------------------|----------------------------------------|----------------------------------------|---------------------|
| Bockeria et al., 2007 | 29                | 20                                     | 9                                      | 5                   |
| Bockeria et al., 2013 | 29                | 17                                     | 12                                     | 7                   |
| Trivedi et al., 2006* | 45                | 31                                     | 14                                     | 20                  |
| Zhang et al., 1997  | 24                | 24                                     | Nil                                    | 8                   |
| Our study 2020     | 60                | 53                                     | 7                                      | 7.5                 |

*This study included the surgeries undertaken for isolated UAPA too. UAPA: Unilateral absence of pulmonary artery

Figure 4: The distribution of the follow up data of patients with unilateral absence of pulmonary artery with congenital heart diseases treated at our center and were followed up (n = 21)
follow-up data of Zhang et al, where they followed up 21 out of 24 patients for an average of 7.5 years and all their patients were asymptomatic and saturating well.\[10\]

Our study brought out an interesting distribution of 24 (32.4%) patients of Group II [Table 1] with increased pulmonary flow physiology, which has not been described earlier. They were offered repair of intracardiac shunts so as to establish normal blood supply at sub systemic pressures to at least one lung via the main pulmonary artery. In UAPA, the lung having unrestricted blood supply from the main pulmonary artery develops pulmonary arterial hypertension in due course of time, irrespective of the presence or absence of intracardiac shunts.\[11\] The presence of MAPCAs to the ipsilateral lung would also contribute to PAH before and after correction. A follow-up cardiac catheterization would have revealed the progression of PAH in these patients. However, none of the operated patients underwent a repeat cardiac catheterization due to various socioeconomic factors. This however, does not reflect the true natural or modified history of this anomaly owing to the sizable loss to follow-up.

Limitation of the study

This study consists of retrospective descriptive data of a rare anomaly from hospital records over a span of 13 years; thus, uniformity in maintaining the data could not be ensured. The evaluation of the patients was tailored on a case to case basis based on cumulative experience, available diagnostic modalities, and existing scientific knowledge. The measurements of the pulmonary artery could not be retrieved in all patients of Group I. Finally, the loss to follow-up was high. The rarity of the anomaly, the varied socioeconomic backgrounds of the patients and the period spanning almost 20 years poses a huge challenge to follow-up these patients, especially in a resource-constrained setting.

CONCLUSIONS

Our study is one of the largest series of UAPA with non-unifocalizable MAPCAs associated with other CHDs. This Indian study corroborates with other studies which demonstrated UAPA as most commonly associated with TOF and its variants. A systematic approach to these patients aids surgical management. Once diagnosed, in the vast majority, single-lung corrective surgery is possible, and single lung repairs have favorable long-term outcomes.

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Conflicts of interest
There are no conflicts of interest.

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