Short Communication

Diagnosis and Management of 60 Children with Congenital Vascular Rings: A 10-year Experience

Guo-Qing Fang[1,2], Jing Li[1], Feng Xu[1], Yue-Qiang Fu[1], Ying-Fu Chen[1], Xiao-Juan Ji[3], He-Lin Zheng[4], Si-Si Chen[5]

[1]Department of PICU, Children's Hospital of Chongqing Medical University, Chongqing 400014, China
[2]Department of Pediatrics, The Ninth People's Hospital of Chongqing, Chongqing 400700, China
[3]Department of Cardiology, Children's Hospital of Chongqing Medical University, Chongqing 400014, China
[4]Department of Radiology, Children's Hospital of Chongqing Medical University, Chongqing 400014, China
[5]Department of Respiratory, Children's Hospital of Chongqing Medical University, Chongqing 400014, China

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Introduction

Congenital vascular rings (VRs) are a group of rare cardiovascular anomalies that result from abnormal embryonic development of the paired aortic arches or branching pulmonary arteries. Clinical presentation of VR depends on its anatomy, degree, and site of compression. Stridor, recurrent upper respiratory tract infections and feeding difficulty were frequent symptoms of VR. Barium swallow test, echocardiography, computed tomography (CT) and magnetic resonance imaging (MRI) present as effective diagnostic tools for VR evaluation. Surgical interventions are required for symptomatic VR patients, and result in favorable outcomes in most cases. However, tracheal stenosis (TS) can often present and be the cause of surgical failure.

In this retrospective study, data from 60 cases of congenital VR in the Children's Hospital of Chongqing Medical University were presented, with the aim of analyzing the clinical characteristics, relative values, advantages and disadvantages of various methods used in VR diagnosis, as well as results of surgical treatment and follow-up assessment.

Methods

Medical records were reviewed from 60 patients with congenital VR. Demographic data included: Sex, age at diagnosis, presenting symptoms, diagnostic tools, and diagnosed subtypes. Patients were assigned into four VR subtypes: (1) Double aortic arch (DAA), (2) pulmonary artery sling (PAS), (3) right aortic arch with a persistent left ligamentum arteriosum (RALL), and (4) others. Cases were evaluated by echocardiography, CT angiography, barium esophagogram and fiberoptic bronchoscopy as appropriate. Management included surgical and nonsurgical treatments. In cases of surgical operation, data regarding age at operation, duration of airway intubation, duration of intensive care unit (ICU) stay, and postoperative complications were analyzed. As for mortality cases, data included the interval between diagnosis and operation, cause of death, and other risk factors such as age at presentation/death and VR type.

Statistical analysis

Data are presented as medians with ranges, frequencies and means with standard deviations as appropriate. Continuous variables and categorical data were compared using nonparametric Rank-sum test and Fisher's exact test, respectively. Analyses were done via the Statistics Analysis System (SAS, SAS Institute Inc., USA). A $P < 0.05$ was considered as statistically significant.

Results

Characteristics of patients

Sixty patients (40 males and 20 females) were enrolled into the study, with a median age of 7.65 months (range: 1 day to 6 years and a month) at presentation. According to the different VR subtypes, PAS occurred in 25 patients (41.67%), DAA in 21 patients (35%), RALL in 12 patients (20%), and other types in 2 patients (3.33%). In most of the 60 cases, respiratory symptoms were commonly presented, recurrent pulmonary infections, stridor and dyspnea were encountered in 52 cases (86.67%). Gastrointestinal symptoms, such as vomiting were noted in two infants (3.33%). Two patients (3.33%) were found to be asymptomatic at the time of diagnosis. Overall, 30 patients (50%) showed VR symptoms during the neonatal period, and 12 patients (20%) presented at birth.

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Address for correspondence: Dr. Jing Li, Department of PICU, The Children's Hospital of Chongqing Medical University, Chongqing 400014, China
E-Mail: lijingwangyi@126.com

Diagnostic methods of congenital vascular rings

All cases were evaluated by echocardiography and demonstrated VRs in 32 patients (53.33%). CT angiography was performed in 55 patients, which demonstrated VRs in 53 patients (96.36%) [Figure 1]. Barium esophagogram was performed in 12 patients, and fiberoptic bronchoscopy was performed in 10 patients while cardiac catheterization was performed in one patient. Our findings revealed that barium esophagogram and fiberoptic bronchoscopy had a diagnostic rate of 83.33% and 90%, respectively, while cardiac catheterization was 100%.

Associated anomalies

Cardiovascular associated anomalies were present in 28 patients (46.67%) including atrioventricular (atrial septal defect) and ventricular septal defects. Complex congenital heart disease was found in seven patients including pulmonary atresia, interrupted aortic arch, transposition of the great arteries, and persistent truncus arteriosus.

Noncardiac associated anomalies mainly included airway constriction in 45 patients (75%), esophageal stricture in 10 patients (16.67%), and both in nine patients (15%).

Surgical management

Forty patients (66.67%) underwent surgical repair at a median age of 8.17 months (range: 1.7 months to 6.17 years). The median time for extracorporeal circulation procedures was 77 min (range: 49–335 min) and the median aortic cross clamp time was 37 min (range: 22–115 min).

Fifteen patients with PAS underwent division of the left pulmonary artery with re-implantation to the main pulmonary artery through median sternotomy. Twelve cases with DAA were repaired through left posterolateral thoracotomy or median sternotomy. Surgical division of the nondominant arch and mobilization of the VR was performed. Twelve patients with RALL underwent division of the ligamentum arteriosum through left posterolateral thoracotomy or median sternotomy. Occlusion of the left subclavian artery was performed in one case. In addition, tracheoplasty was performed in two patients among all the 40 cases underwent surgical management.

The median ICU stay after surgery was 48 h (range: 10 h to 74 days). The median ICU stay for patients with PAS was relatively longer than non-PAS patients, which was 72 h (range: 10 h to 46 days) versus 21 h (range: 15 h to 74 days), respectively. The median time of ventilator use after surgery was 21 h (range: 2 h to 46 days). The median duration of ventilator usage was also longer in patients with PAS versus other types, which was 30 h (range: 4 h to 46 days) versus 21 h (range: 2 h to 10 days), respectively.

Postoperative complications included: Respiratory failure (six cases), chylothorax (four cases), atelectasis (two cases), pneumonitis (three cases), acute respiratory distress syndrome (one case), vocal cord palsy (one case) and sepsis (one case).

Surgical repair was not performed in 20 patients. One patient remained asymptomatic, and one PAS patient died of respiratory failure and pulmonary infection. Ten patients who had complex anomalies were discharged and lost-in-follow-up.

Mortality

There were seven mortalities (11.67%) in 60 cases. One patient with PAS died of pulmonary infection and respiratory failure before surgical management. There were six mortalities within the 40 patients who underwent surgical management. One patient of PAS associated with ring-sling syndrome died intraoperatively due to cardiac dysfunction. Five patients died after surgical management, which included three cases of PAS, one case of DAA and one case of RALL.

Statistical analysis [Table 1] showed that the survival group and the death group were significantly different in terms of the presence of VR associated with complete tracheal cartilage rings ($P = 0.002$).

Discussion

A VR is a rare anomaly of the great arteries, which could compress the trachea or esophagus, and usually becomes symptomatic in infancy or early childhood. The subtype distribution in this review was mainly comprised of PAS (41.67%) and DAA (35%). Similar to another retrospective review in Korea,[1] 10 patients (28.6%) presented with DAA, while eight patients (22.9%) presented with PAS, indicating a DAA and PAS dominant subtype distribution in both areas. In contrast, studies in US[2] and Turkey[3] suggested a higher scale of RALL (42%) and left

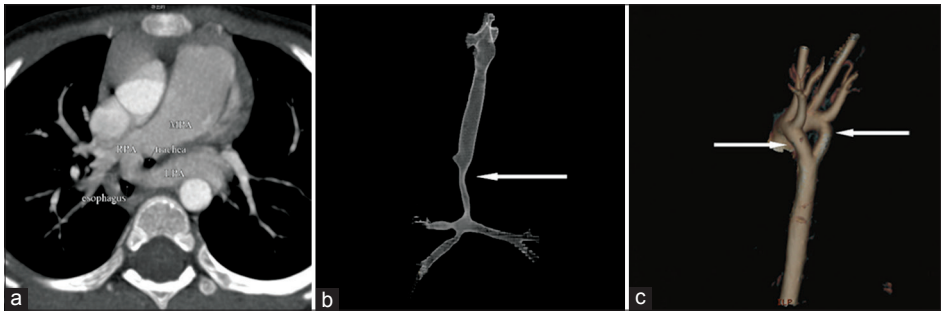

Figure 1: Three-dimensional structures of diverse vascular ring subtypes when reconstructed from cardiac computed tomography angiography. (a) Pulmonary artery sling (PAS): The origin of the left pulmonary artery from the right pulmonary artery; (b) PAS: Left pulmonary artery compression at the bronchus (white bold arrows); (c) Double aortic arch: Two aortic arches (white bold arrows) comprise the complete ring in this patient.

Table 1: Comparison of clinical data between the death group and survival group in congenital VR surgical cases

Variables	Death (n=6)	Survival (n=34)	P
Type			
RALL	1	11	0.648
DAA	1	11	0.648
PAS	4	11	0.174
Weight (g)	2591.7 ± 657.6	2942.1 ± 586.7	0.193
Female	2	11	1.00
Age at diagnosis (months)	5.5 (3.0–8.0)	8 (5–18)	0.133
Age at operation (months)	7.5 (4.0–8.0)	9 (6–19)	0.232
Cardiac anomalies	2	16	0.673
Complex cardiac anomalies	2	3	0.154
Tracheal stenosis	5	24	1.00
Long-distant tracheal stenosis	3	8	0.319
Complete tracheal cartilage rings	3	0	0.002
Time of respirator usage	98 (23–1104)	14.5 (6.0–39.0)	0.078
ICU stay (h)	240 (28–552)	23 (21–168)	0.225
Interval between presence and diagnosis (months)	4 (2–7)	5.5 (2.0–16.0)	0.436

RALL: Right aortic arch with persistent left ligamentum; PAS: Pulmonary artery sling; DAA: Double aortic arch; ICU: Intensive care unit; VR: Vascular rings.

aortic arch with aberrant retroesophageal right subclavian artery (56.8%), respectively. They reported the scale of patients with PAS were only 4% and 0.05%, respectively, which showed the significant heterogeneity of VR subtypes in these patient populations. We hypothesize that among the VR patients, the subtype incidence of PAS may vary from districts and ethnic populations as various customs could potentially affect risk factors of congenital VR morbidity. These data suggest that the PAS susceptibility might be higher in the Asian region when compared with European and American regions.

Common presenting symptoms of VR include respiratory and gastrointestinal symptoms. The primary clinical presentation for VR in our study was respiratory symptoms (87%), which included severe dyspnea, stridor and repeated respiratory tract infections. Nearly half of the patients showed symptoms in the neonatal period, with 12 cases presenting symptoms at birth particularly and only two patients (3.3%) harboring an asymptomatic diagnosis. However, previous studies[1] suggested that the proportion of asymptomatic patients was significantly higher, with 11 asymptomatic cases (31%). Respiratory symptoms are the dominant clinical presentation in patients with VR of different subtypes; however, our study highlights that more cases have severe symptoms and higher clinical morbidity. This could be due to the large proportion of patients with PAS and DAA.

Early diagnosis is key to congenital VR management since VR shares clinical symptoms with other respiratory disorders, it can be easily misdiagnosed if there is lack of adequate recognition of symptoms. Thus, relevant imaging tests are necessary for auxiliary accuracy of VR diagnosis. Barium esophagram was recommended as a primary diagnostic method in previous years; however, development of CT and MRI technology has improved patient diagnostics and thus, barium esophagram is no longer required before proceeding with VR repair. In this study, we performed CT angiography in 55 patients to accurately diagnose VR. The diagnostic coincidence rate of this technique was shown to be approximately 96.36%. Prior studies have demonstrated that CT angiography could extract anatomical and limited functional data within a short period without the need for pharmacological intervention and could be used for quick and accurate VR diagnosis.[4] MRI is superior in its evaluation of cardiac anatomy and physiology when compared to CT angiography; however, it requires significantly prolonged usage time and management since it requires sedation or general anesthesia in young children. As a result, MRI was not widely applied for VR diagnosis in our study. Thus, we recommend CT angiography as a capital option for VR diagnosis, in spatial relation to the trachea and esophagus. On the other hand, the diagnostic coincidence rate of echocardiography was only 53.33%, which demonstrated the limitation of echocardiography for accurate VR diagnosis. Regardless, this protocol could still provide information on other observed cardiac anomalies in VR patients, which could assist with VR diagnosis. We also performed fiberoptic bronchoscopy in 10 patients harboring TS. It has been reported that bronchoscopy can help visualize the external, pulsating mass compressing on the trachea in most cases. Our study revealed that it was superior in assessing the endoluminal airway and vocal fold mobility, as well as in diagnosing associated airway lesions.

Operative interventions are often required urgently to treat VR-related symptoms with timely surgeries being paramount in symptomatic patients. Surgical treatments were conducted on 40 patients in our study, with six mortalities occurring within patients who underwent operations, and four cases with PAS of which had significantly prolonged ICU stay. Previous studies have indicated VR associated with complex cardiac diseases show poor prognosis.[1] Nevertheless, we found that the poor prognosis in the most complex patients with congenital VR was associated with complete tracheal cartilage rings (P = 0.002). Similarly, Chiu *et al*. suggested that VR repair in patients with tracheal narrowing would carry a higher risk of mortality compared to patients undergoing VR repair in the absence of symptomatic airway obstruction.[5] Therefore, we highlighted that severe TS and in particular, complete tracheal cartilage rings, could be a high-risk factor for poor prognosis in congenital VR patients. However, there are no effective standards of management for surgical interventions and it remains obscure whether tracheal repair operations and conservative treatments are important in decreasing patient mortality.[5] It is also suggested that not all congenital VR patients with underlying airway obstructions require combined tracheal repair, thus, children might benefit from conservative treatments, in some circumstances. We highlighted two VR cases that underwent tracheoplasty; however, one patient

died intra-operatively while the other died postoperatively. These data demonstrate that the simultaneous reconstruction of congenital VR and severe TS is a challenge for surgical intervention. Future studies evaluating the management of congenital VR patients with TS are still required.

Infantile patients presenting with repeated respiratory symptoms have a predisposition to congenital VR, particularly when medical treatments are ineffective. The susceptibility of PAS might be higher in the Asian region when compared to European and American regions. CT angiography presents as an optimal method to validate VR diagnosis. VR combined with complete tracheal cartilage rings is a risk factor for mortality.

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