Editorial

Editorial on "vascular ring diagnosis and management: notable trends over 25 years"

Jill J. Savla, Paul M. Weinberg

Children's Hospital of Philadelphia, Division of Cardiology, Philadelphia, PA, USA

Correspondence to: Paul M. Weinberg, MD. Children's Hospital of Philadelphia, 3401 Civic Center Blvd, Philadelphia, PA 19104, USA.
Email: weinberg@email.chop.edu.

Provenance: This is a Guest Editorial commissioned by the Section Editor Xicheng Deng (Department of Cardiothoracic Surgery, Hunan Children's Hospital, Changsha, China).

Comment on: Evans WN, Acherman RJ, Ciccolo ML, *et al.* Vascular Ring Diagnosis and Management: Notable Trends Over 25 Years. World J Pediatr Congenit Heart Surg 2016;7:717-20.

Submitted Feb 28, 2017. Accepted for publication Mar 01, 2017.
doi: 10.21037/tp.2017.03.05
View this article at: http://dx.doi.org/10.21037/tp.2017.03.05

Vascular rings occur due to some perturbation in the normal sequence of development and selective involution of paired aortic arches to form a definitive left or right aortic arch. The two most common types of vascular rings are double aortic arch (DAA) and right aortic arch with an aberrant left subclavian artery and a left patent ductus arteriosus or ligamentum arteriosum (RAA-ALS) (1,2). The prevalence of vascular rings is unknown because many patients remain asymptomatic and are undiagnosed.

A recently published study by Evans and colleagues reviewed the diagnosis and management of vascular rings before and after the introduction of prenatal diagnosis (3). In Southern Nevada from 1990 to 2015, they diagnosed 75 patients with an isolated vascular ring and 17 patients with a vascular ring in addition to significant congenital heart disease. Of the 75 patients with an isolated vascular ring, 58 patients (77%) had RAA-ALS, 15 patients (20%) had DAA, and 2 patients (3%) had a pulmonary artery sling. Pulmonary artery sling, which is the anomalous origin of the left pulmonary artery from the right pulmonary artery, is technically not a true vascular ring since it does not encase both the trachea and the esophagus.

As indicated in the study, after the three-vessel fetal echocardiography view was introduced in 2004, the rate of prenatal detection increased significantly from 0% before 2006, to 55% in 2010, and then up to 70% prenatal detection of diagnosed vascular rings since 2013 (3). Compared to postnatal transthoracic echocardiography, vascular rings are theoretically easier to recognize by fetal echocardiography because the ductus arteriosus is patent, and the paucity of air in the fetal lungs allows for a better visualization of thoracic structures. However, it can be challenging to determine some details by fetal echocardiography, such as which side of a DAA will be the dominant side. Prenatal diagnosis also differs from postnatal diagnosis in the fact that the fetus is typically asymptomatic and the findings may be incidental. This highlights an interesting possibility that the distribution of vascular ring types in fetuses may be different than our previous understanding of vascular ring types, which was historically based on symptomatic patients referred for surgical repair.

Vascular rings due to DAA frequently become symptomatic because the ring around the trachea and esophagus is often tight. Therefore, it is understandable why published series of surgical patients described DAA as the most common type of vascular ring (4-6). In contrast, vascular rings due to RAA-ALS can be asymptomatic or only have mild symptoms because the ring is relatively loose. Interestingly, of the 75 patients in Evans' study with an isolated vascular ring, 23 patients were still asymptomatic at the time of the publication and all 23 had RAA-ALS (3).

More recent articles that focus on prenatal diagnosis support the notion that RAA-ALS may be more prevalent than DAA in fetuses and asymptomatic patients, who previously would never have been diagnosed (7). A study by Galindo *et al.*, analyzed 18 patients with a prenatal diagnosis

of vascular ring, 15 of which had RAA-ALS and 3 of which had DAA (8). There were 16 live births and 15 of those patients remained completely asymptomatic, with a mean postnatal follow-up time of 31 months. One patient had mild symptoms but did not require surgical repair.

Another study by Razon *et al.*, followed 58 patients with a fetal diagnosis of right aortic arch (with or without a vascular ring) and normal or near-normal intracardiac anatomy (9). Their postnatal cohort consisted of 23 patients with RAA-ALS and 8 patients with DAA. With a mean postnatal follow-up time of 49 months, only 11 patients developed symptoms. Of these 11 symptomatic patients, 8 had DAA, 2 had RAA-ALS, and 1 had right aortic arch with mirror-image branching with a left ligamentum arteriosum arising directly from the descending thoracic aorta (retroesophageal ligamentum). Ten of these 11 symptomatic patients underwent surgical repair and 1 patient with RAA-ALS did not require surgery because their symptoms resolved. This study demonstrates that although RAA-ALS may be more prevalent among fetuses, patients with DAA still tend to develop symptoms more often.

Evans and colleagues reported that of the 52 patients with an isolated vascular ring who underwent surgical repair, 9 patients had surgery in the 15-year period from 1990 to 2005 and 43 patients had surgery in the 10-year period from 2006 to 2015 (3). During the earlier surgical era, 6 of the 9 patients (67%) had DAA, while 9 of the 43 patients (21%) during the later surgical era had DAA. Based on these results, the authors concluded that the percentage of patients with RAA-ALS has increased in Southern Nevada over the past 25 years. The authors also concluded that the rate of prenatal diagnosis has increased and that patient age at surgical repair of an isolated vascular ring has decreased. However, it is unlikely that the actual incidence of RAA-ALS compared to DAA has changed, but rather that the onset of prenatal diagnosis has influenced clinical practice patterns regarding referral for surgery.

Clinicians taking care of patients with a prenatal diagnosis of a vascular ring may inherently have a higher index of suspicion that new respiratory symptoms (i.e., wheezing, cough, stridor) or gastrointestinal symptoms (i.e., dysphagia, vomiting) are related to the vascular ring, instead of more common pediatric diagnoses such as asthma or reflux. Prenatal diagnosis is beneficial for patients who truly become symptomatic from their vascular ring, in order to ensure a timely surgical repair. However, we must also consider that as we increase the rate of prenatal detection and identify potentially asymptomatic patients with vascular rings, we may be reacting sooner and operating on less symptomatic patients.

This is especially true given the natural history of vascular rings. A study of 11 unoperated patients with vascular rings and mild symptoms at the time of diagnosis, reported that 9 patients were entirely free of their symptoms by 1-2 years after the diagnosis (10). Only 2 of the 11 patients had persistent symptoms, with a median follow-up time of 7 years. This study was the basis for the historically conservative stance on surgery in patients with no symptoms or only mild symptoms, because they may outgrow their symptoms in early childhood. A more recent case series of asymptomatic patients also recommends watchful waiting with clinical observation of asymptomatic vascular rings (11).

When the decision is made to refer a patient with significant or persistent symptoms for surgical repair of a vascular ring, preoperative imaging with either magnetic resonance imaging (MRI) or computed tomography (CT) is essential to provide an accurate and complete diagnosis (12). For example, identifying the dominant side of a DAA or evaluating for distal arch obstruction can change the surgical approach of a lateral thoracotomy.

In conclusion, the incidence of RAA-ALS compared to DAA is unlikely to have changed, but RAA-ALS may be more prevalent in asymptomatic patients than was previously known. The introduction of prenatal diagnosis for vascular rings has influenced clinical practice because we can identify patients who remain asymptomatic or only have mild transient symptoms. This is beneficial for some patients who truly become symptomatic and results in a timely surgical repair. However, a fetal diagnosis of vascular ring should not be the only indication for surgery. Although cardiothoracic surgery has become much safer in the current era, it is still not entirely risk-free, and lateral thoracotomy can sometimes result in scoliosis. Therefore, the clinician should carefully weigh the risks and benefits of operating on patients with asymptomatic vascular rings.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

References

1. Backer CL, Mongé MC, Popescu AR, et al. Vascular rings. Semin Pediatr Surg 2016;25:165-75.
2. Juraszek AL, Guleserian KJ. Common aortic arch anomalies: diagnosis and management. Curr Treat Options Cardiovasc Med 2006;8:414-8.
3. Evans WN, Acherman RJ, Ciccolo ML, et al. Vascular Ring Diagnosis and Management: Notable Trends Over 25 Years. World J Pediatr Congenit Heart Surg 2016;7:717-20.
4. Roesler M, De Leval M, Chrispin A, et al. Surgical management of vascular ring. Ann Surg 1983;197:139-46.
5. van Son JA, Julsrud PR, Hagler DJ, et al. Surgical treatment of vascular rings: the Mayo Clinic experience. Mayo Clin Proc 1993;68:1056-63.
6. Anand R, Dooley KJ, Williams WH, et al. Follow-up of surgical correction of vascular anomalies causing tracheobronchial compression. Pediatr Cardiol 1994;15:58-61.
7. Patel CR, Lane JR, Spector ML, et al. Fetal echocardiographic diagnosis of vascular rings. J Ultrasound Med 2006;25:251-7.
8. Galindo A, Nieto O, Nieto MT, et al. Prenatal diagnosis of right aortic arch: associated findings, pregnancy outcome, and clinical significance of vascular rings. Prenat Diagn 2009;29:975-81.
9. Razon Y, Berant M, Fogelman R, et al. Prenatal diagnosis and outcome of right aortic arch without significant intracardiac anomaly. J Am Soc Echocardiogr 2014;27:1352-8.
10. Godtfredsen J, Wennevold A, Efsen F, et al. Natural history of vascular ring with clinical manifestations. A follow-up study of eleven unoperated cases. Scand J Thorac Cardiovasc Surg 1977;11:75-7.
11. Loomba RS. Natural History of Asymptomatic and Unrepaired Vascular Rings: Is Watchful Waiting a Viable Option? A New Case and Review of Previously Reported Cases. Children (Basel) 2016;3. pii: E44.
12. Smith BM, Lu JC, Dorfman AL, et al. Rings and slings revisited. Magn Reson Imaging Clin N Am 2015;23:127-35.

Cite this article as: Savla JJ, Weinberg PM. Editorial on "vascular ring diagnosis and management: notable trends over 25 years". Transl Pediatr 2017;6(2):83-85. doi: 10.21037/tp.2017.03.05