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Non-invasive diagnosis of aortic arch anomalies in children – 15 years of own experience

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echocardiography, right aortic arch, double aortic arch, Kommerell's diverticulum, aberrant subclavian artery

Abstract

Aim: To summarize our experience in echocardiographic diagnosis of aortic arch anomalies in pediatric patients. **Materials and methods**: A retrospective assessment of echocardiographic findings in Echo-Lab patients of the Pediatric Cardiac Surgery Department, who were diagnosed with an anomalous aortic arch between 2003 and 2018. **Results**: The diagnosis of an abnormal course of the aortic arch and/or its branches was established in 115 children aged between 4 days and 17 years. The following types of anomalies were detected: left aortic arch with aberrant right subclavian artery in 42 patients; right aortic arch with left brachiocephalic trunk in 14 patients; right aortic arch, aberrant left subclavian artery in 36 patients; double aortic arch in 14 patients; and other, more complex types in 9 patients. The main elements of defects were correctly identified by echo in all cases. We decided to additionally perform computed tomography angiography in 32 patients to clarify all details necessary to qualify patients for surgery and establish the surgical plan. **Conclusions**: 1. Echocardiography strictly following the pre-determined protocol has 100% sensitivity in the detection of basic elements of aortic arch anomaly and is a perfect tool for diagnostic process initiation. 2. Since it is not possible to visualize all anatomic details, the qualification for surgery should by based on computed tomography angiography or cardiac magnetic resonance imaging, which precisely visualize both abnormal vessels and compressed structures.

Introduction

Aortic arch anomalies are a diverse group of congenital anatomical defects of the main artery, which may be the primary cause of significant symptoms[1–3]. When coexisting with other congenital cardiovascular, respiratory and gastrointestinal defects, they affect both their symptoms and the treatment process.

Abnormal spatial relations between the aortic arch and its branches and the esophagus and trachea in patients with vascular rings may cause respiratory symptoms and dysphagia. Furthermore, there are clinical situations where different, initially asymptomatic courses of the aortic arch may significantly hamper surgical treatment, e.g. congenital esophageal or upper respiratory defects.

It is natural that patients reporting to an echocardiographic laboratory in a multidisciplinary pediatric teaching hospital with departments of neonatal pathology, pulmonology and gastroenterology include children suspected of vascular abnormalities requiring thorough diagnosis. Patients from cardiac departments, which also feature echo laboratories, represent a large proportion of pediatric patients with vascular malformations. The diagnostic process includes cases of patients with previously diagnosed vascular ring, who are referred for surgical treatment, as well as those diagnosed during a thorough qualification for surgery due to other congenital heart defects.

Materials and methods

Echocardiography is of key importance for the diagnosis of anatomical abnormalities of the main artery in our clinical practice despite limitations associated with its thoracic location and the fact that a significant part of its course is obscured by the lungs, airways and osseous structures. For the purpose of the study, we conducted a retrospective analysis of medical records of 115 pediatric patients who were ultrasonographically diagnosed with aortic arch abnormalities known as vascular rings. Patients with aortic arch pathologies such as aortic coarctation, supravalvular aortic stenosis or interrupted aortic arch were excluded from the study.

We analyzed clinical data, echo recordings, as well as data on the treatment of 115 children with aortic arch abnormalities. The group included 58 boys and 57 girls aged between 1 day and 17 years (full pediatric age range).

Results

The following aortic arch anomalies were detected in the study group:

- Left aortic arch (LAA) + aberrant right subclavian artery (ARSA);
- Right aortic arch (RAA) + left brachiocephalic trunk (LBCT);
- RAA + aberrant left subclavian artery (ALSA);
- double aortic arch (descending aorta, DAoA); and
- untypical, rare variants (right aortic arch with isolated left subclavian artery, right aortic arch with ductus arteriosus from the descending aorta to the right pulmonary artery, right aortic arch with an aberrant left subclavian artery and ductus arteriosus between the aorta and the left pulmonary artery, right aortic arch with the left descending aorta, left aortic arch with the right descending aorta and ARSA).

Each of the above mentioned subgroups was assessed for the vascular morphology of the aortic arch, number, sex distribution, coexisting congenital heart defects, dominant clinical manifestations, as well as the type of diagnostic method which specified the final diagnosis in a sufficient way to make a decision on potential surgical treatment. The data obtained are summarized in Table 1.

Discussion

Echocardiography was performed in all patients in the study group to assess the location and course of the aortic arch, as well as the arrangement and course of aortic arch branches and their spatial relationship with the esophagus and the trachea. Echocardiography initiated diagnostic imaging in a vast majority of patients diagnosed based on the assessment of symptoms (pulmonary, gastrointestinal or laryngological) suggestive of an abnormal course of the aorta and its branches, patients diagnosed accidentally during echo due to other reasons, as well as patients qualified for cardiac surgeries. Previous contrast-enhanced esophageal radiography showing characteristic esophageal deformation was reported for only seven children with respiratory and gastrointestinal disorders. A CT angiography, which also revealed aortic arch abnormalities, was performed (in another center) as part of the diagnostic process in 4 children with congenital heart defects.

Echocardiography showed 100% sensitivity in the assessment of the basic anatomy of a single arch[4–8]. Aortic arch location and the course of its main branches were accurately identified in all cases where echo preceded CT angiography. Therefore, we decided not to perform esophageal radiography[2,9] and resigned from CT angiography is the case of absence of disturbing symptoms as the next step of the management in our laboratory.

Echocardiography also showed 100% sensitivity in the diagnosis of double aortic arch with both patent branches with similar diameters[2]. Echocardiography showed lower diagnostic accuracy in patients with double aortic arch and atresia of one of its branches. Atresia of the left branch segment located between the left subclavian artery and the

Tab. 1. *Characteristics of the study group in terms of the number of different types of aortic arch anomaly, the main clinical manifestations and the method used for definitive cardiac surgical diagnosis*

Type of anomaly	*N*	♂	♀	CHD	Dysphagia	Respiratory symptoms	Echo-based diagnosis	CT-based diagnosis
LAA + ARSA	42	21	21	12	9	10	30	12
RAA + LBCT	14	9	5	14	–	–	14	
RAA + ALSA	36	20	16	20	12	11	36	
DAoA	14	7	7	2	13	13*		14
Other	9	1	8	2	5	6	3	6
Total	115	58	57	50	9	23	83	32

* Increased severity of respiratory symptoms; CHD – congenital heart disease requiring cardiac surgery, LAA – left aortic arch; ARSA – aberrant right subclavian artery; RAA – right aortic arch; LBCT – left brachiocephalic trunk; ALSA – aberrant left subclavian artery; DAoA – double aortic arch (descending aorta)

descending aorta was detected in each of the seven analyzed cases. The echocardiographic image was similar to the one seen in the right aortic arch with left brachiocephalic trunk. Although a more arched, posteriorly directed course of the patent segment of the double aortic arch was helpful in the differentiation[7,8], significant diversity in the shape of the brachiocephalic trunk and the patent segment of the arch was observed in both types of anomaly. The absence of vascular ring in RAA + LBCT, and hence the lack of compressive symptoms in these patients as opposed to the double aortic arch, is the basic clinical difference between these forms of aortic arch anomaly. Furthermore, we identified RAA + LTBC almost exclusively in children in the group of conotruncal malformations (tetralogy of Fallot; pulmonary atresia with ventricular septal defect, PA/VSD; double outlet right ventricle with ventricular septal defect, DORV/VSD)[6,7]; therefore, the lack of such an association may also be a diagnostic hint in the case of doubts[2,10]. CT angiography was performed in all children with echocardiographically suspected double aortic arch in order to obtain a more precise anatomical picture. Although CT angiography is unable to show an obstructed aortic segment, the three-dimensional vascular reconstruction allows for a much more suggestive reconstruction of their shape[8] and is more likely to show the presence of a blind segment compared to echo. Ultimately, direct surgical visualization of the aortic segment undetectable by diagnostic imaging is the only reliable diagnostic method.

Patients with atypical anomalies, right aortic arch and isolated left subclavian artery in particular, are another subgroup in which no definite diagnosis was made based on echocardiography[11]. A first narrow branch of the right aortic arch without typical division, i.e. the left common carotid artery, was visualized using echo in each of the three cases in the analyzed material. In the vast majority of cases, such an image is characteristic for an aberrant left subclavian artery (LSA); however, we did not visualize the proximal segment of this artery in any of our patients. However, the proximal (pulmonary) part of the patent ductus arteriosus passing upward and leftward, hence most likely arising from the subclavian artery, was visualized. The entire picture raised a suspicion of LSA isolation. CT angiography, which clarified the diagnosis, was performed in one of the patients, who was prepared for a correction of the tetralogy of Fallot. The other patient later underwent a correction of a complete AV defect; however, no vascular malformations were diagnosed at that time. Such a suspicion was raised during later follow-up and confirmed based on nuclear magnetic resonance (NMR). No intracardiac defect was found in the third patient in this subgroup. Further diagnosis was planned after the outpatient echocardiography; however, the child did not report for computed tomography. CT angiography (4 patients) and NMR (1 patient) were also performed in patients with right aortic arch and left ductus arteriosus from the descending aorta and, finally[12], an encircling aortic arch[13], to resolve the doubts.

It is generally assumed that symptoms of esophageal and tracheal compression due to an abnormally coursing vessel are an indication for surgical correction of a vascular ring[9,13–15]. For this reason, the aim of the diagnosis is not only to show the presence of abnormal vessels, but to also confirm that their course causes distortion of the esophagus and upper respiratory tract, which may lead to symptoms. Although ultrasonography usually allows for visualization of the trachea and the esophagus (in small children), as well as shows their distortion in some cases, these images are not clear enough to be a basis for therapeutic decisions. Therefore, when qualifying patients with vascular rings, we rely on techniques which, in addition to vessels, visualize the esophagus and the respiratory tract, such as CT, NMR, and endoscopy in the case of doubts[16–20]. Patients with congenital intracardiac defects, when the surgery of vascular ring involves ductus/ligamentum arteriosus division, e.g. the right aortic arch with ductus arteriosus passing from the left pulmonary artery to the brachiocephalic trunk or subclavian artery, are an exception. If such an anomaly is identified and sufficient data on intracardiac surgical extent is obtained using ultrasonography, further extension of the diagnosis is not needed for making therapeutic decisions.

Conclusions

1. Echocardiography is sufficient to identify and precisely define most anatomical elements of an abnormal aortic arch and a starting point for the planning and implementation of other imaging modalities in doubtful cases.
2. Extended diagnosis allowing for three-dimensional reconstruction of vessels, airways and the esophagus is very helpful in determining indications for surgical treatment and planning surgical extent.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

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