The Egyptian Journal of Radiology and Nuclear Medicine xxx (2017) xxx–xxx

Contents lists available at ScienceDirect

The Egyptian Journal of Radiology and Nuclear Medicine

journal homepage: www.sciencedirect.com/locate/ejrnm

Original Article

Congenital biscuspid aortic valve in pediatric and early adults: Does valvular phenotype affect other parameters?

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ARTICLE INFO

Article history:
Received 2 February 2017
Accepted 3 July 2017
Available online xxxx

Keywords:
Bicuspid aortic valve
Phenotype
Aortic coarctation
Pediatrics

ABSTRACT

Aim of the study: Our study is a prospective study aiming to assess congenital bicuspid aortic valve using cardiac MRI and to detect a relationship between the leaflet fusion pattern and other functional parameters including valvular regurge, stenosis and pressure gradient.

Patients and methods: This prospective study included 114 cases 104 of them had bicuspid aortic valve, diagnosed by echocardiography while the rest (10 cases) were normal control cases. All functional and morphological cardiovascular abnormalities were recorded as well as any associated congenital diseases. Two different radiologists read the MRI blindly to each other.

Results: Patients age ranged from 40 days up to 20 years with 62 patients being in the pediatric age group (<12 years). They were 78males and 26 females.

We found 54 cases (53.8%) of 1-RL morphology, 34 cases (32.7%) of 1-RN morphology, 8 cases (7.7%) of 0-AP and 6 cases (5.8%) of 0-Lateral. Aortic stenosis and regurgitation were found in 80.75% of cases. Aortic stenosis was the most common valvular lesion being more evident in the 1-RN valve type. Aortic regurge was more predominant in the pediatric age group with no predilection for a specific valve phenotype. Left ventricular function was normal in most of our cases with only 10 cases showing impairment due to long standing valvular lesion.

Thirty-eight cases (36.5%) had associated aortic dilatation. Twenty-six patients (25%) showed associated aortic coarctation. Intra cardiac shunts (24 patients 23%) were the second most common associated anomaly after coarctation. Most of the associated congenital anomalies were found with the 1-RL valve morphology type. Other associated congenital anomalies were detected in 44 patients (42.3%).

Conclusion: Cardiac Magnetic resonance should be performed for patients with bicuspid aortic valve to determine the valve phenotype, function and aortopathy for risk stratification and surgical management. Patients with RL type should be properly imaged to detect associated congenital anomalies.

1. Introduction

Bicuspid aortic valve (BAV) variant is the most common congenital heart disease; with an incidence of 0.5 to 2% of the general population. It is more common in males than in females with a 3:1 ratio [1].

Individuals with BAV are more liable to valvular disease earlier than those with tricuspid aortic valve including aortic stenosis (AS), regurge (AR) [2] and infective endocarditis [1].

The BAV disease is not limited only to the aortic valve but it may affect the aortic artery segments till the arch (root, ascending and arch) as well as all the tissues coming from the neural crest. Many other conditions are associated to the BAV disease as ascending aortic dilatation or aneurysm, coarctation of the aorta [2], supravalvular aortic stenosis (William's syndrome), patent ductus arteriosus, ventricular septal defect, and congenital coronary anomalies [1]. In addition it is considered as a risk factor for acute aortic dissection [3].

Till now the exact pathogenesis of the BAV formation is not fully understood. However, its association with other congenital anomalies enhances the theory of genetic component. Fusion of the aortic cusps during valvu-genesis is the suggested mechanism [4].

Careful assessment of the morphologic patterns of BAV, valve-related complications, and the diameter of the aortic root and

Peer review under responsibility of The Egyptian Society of Radiology and Nuclear Medicine.

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https://doi.org/10.1016/j.ejrnm.2017.07.002
0378-603X/

ascending aorta are of great value in the surgical decision making. In addition BAV patients need continuous follow up to detect and treat any developed conditions thus preventing complications [5].

In the last decade, cardiovascular magnetic resonance (CMR) proved to be a powerful modality for these patients in comparison to the echocardiography for assessment of valvular stenosis (valve area and gradient across the valve), regurgitation (CMR can quantify the regurgitated volume either as an absolute value or as the regurgitated fraction), accurate assessment of ventricular function (the gold standard method), and reliable assessment of the cardiac and vascular anatomy allowing assessment of associated pathologies of the thoracic aorta and other congenital anomalies [6].

We performed this study to properly evaluate the bicuspid aortic valve, its valvular phenotype and associated pathologies which may have certain predilection to the pattern of leaflet fusion.

2. Patients and methods

This prospective study included 114 cases, 104 of them were referred to the radiology department from the pediatric hospital and cardiology department with clinical diagnosis of BAV by echocardiography to evaluate the aortic valve and the associated pathologic conditions through the period from December 2014 till January 2016. Ten normal cases were included as control group. Exclusion criteria included cardiac pacemaker, clipsed cerebral aneurysm, claustrophobia, marked obesity and patients with glomerular filtration rate (GFR) less than 60 ml/min if contrast injection is indicated. Two different radiologists read the MRI blindly to each other.

Human ethics committee approval was obtained from the institutional review board of our institute and have been performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants (patients/guardians) included in the study.

Cardiac MRI was performed for all patients using (Philips-Achieva 1.5 Tesla best the Netherland, or Seimens Aera 1.5 Tesla):

3. CMR imaging protocol

Patients were scanned in supine position using a phased-array cardiac coil (8 channel on the Philips-Achieva 1.5 T, AND 32 channel on the Seimens Aera 1.5 T). Preliminary scout localizers in axial, coronal and sagittal planes were done. Sedation was used for children <6 years. The examination was properly explained to patients >6 yrs with training on breath hold performance.

Images analysis and sequences used:

(1) Assessment of aortic valve morphology and area (planimetry)

We used full stack of in plane images across the aortic valve SSFP/gradient cine [spatial resolution was $1.8 \times 1.8 \times 6$ mm/voxel] to evaluate the valve area, morphology and phenotype. We used Sievers classification [7] for determining the BAV morphological type as well as the presence and position of a raphe.

(2) Assessment of stenosis and regurge

Cine SSFP/gradient LVOT view across the aortic valve and Velocity Encoding Images for the evaluation of *trans*-aortic flow in 30 phase were used. The severity of aortic stenosis was based on the pressure gradient. (<20 mmHg is mild, 20–40 mmHg is moderate and more than 40 mmHg is considered severe). Aortic regurgitation was graded according to Gabriel et al. [8].

(3) Assessment of the left ventricular volumes, function and associated anomalies:

Image acquisition was performed in short axis covering the whole left ventricle and four chamber views using Cine SSFP sequences. The ventricular ejection fraction was considered normal from 55 to 75%, border line: 40–54% and low if <40%. The rest of the volumes was detected.

(4) Assessment of aortic dimension

At the annulus, aortic sinus, sino-tubular junction, ascending aorta, aortic arch and descending aorta using 3D MR whole heart sequence or sagittal HASTE sequence. The aortic diameters were displayed in relation to BSA for each measurement site, and correlated to the Z score for the aortic dimensions.

(5) Assessment of associated myocardial scarring if clinically indicated:

In seven patients known to have long standing aortic stenosis or abnormal wall motion on cine images, delayed enhancement MR imaging was done after intravenous infusion of gadolinium chelate contrast material (0.1–0.2 mmol/kg) followed by a cardiac-gated segmented inversion-recovery–prepared fast gradient-echo sequence 10–15 mins later in short axis and four chamber views.

4. Results

Our study included 104 patients, 26 females (25%) and 78 males ((75%) with their age ranging from 40 days to 20 years (mean age 11.4). The majority (59.6%) of our patients were in the pediatric age group.

Most of our cases (56) showed 1-RN morphology (53.8%), while 34 cases (32.7%) were of 1-RN morphology, 8 cases were 0-AP, 6 cases were 0-lateral. None of our cases had unicaspid or 1-LN types.

Our results showed that 81% of patients with BAV have associated valvular dysfunction with only 19% show normal valvular function. AS was the most common valvular disorder found representing 62 cases (59.6%) with mean age of 18.1, while AR was detected in 48 cases (46%) presenting at younger age with a mean age of 12.3.

We found that most patients with 1-RN phenotype (94.1%) showed valvular dysfunction predominantly AS which represents 82% of the cases. Meanwhile, patients with 1-RL phenotype had lower incidence of valvular dysfunction (75%) with no significant preferences to AS or AR.

According to the severity of aortic stenosis we had 24 patients with moderate AS, 22 with severe AS and 16 showing mild AS. Most of the severe AS patients (16 cases) were of the 1-RN group. Mean pressure gradients was 55 mmHG in 1-RN valve phenotype, 37 mmHg in 1-RL phenotype, 34 mmHg in 0-AP phenotype and 26 mmHg in the 0-lat phenotype.

Different grades of AR were detected in our study ranging from Grade 0 to IV with no association with a specific valve phenotype.

Most of our patients showed normal left ventricular function. The left ventricular ejection fraction in 94 patients (90.4%) were normal, 4 patients (3.8%) were border line and 6 (5.8%) patients showed low ejection fraction denoting heart failure. Left ventricle dilatation was detected in 38 patients (36.5%).

Aortic dilatation was detected in 38 cases. Four of those patients had normal valvular function. Dilatation of the entire ascending aorta including the tubular ascending aorta and sinus of Valsalva (Type A) was the most frequent encountered type of dilatation being detected in 18 cases (47.4%). Isolated dilatation of the

Please cite this article in press as: Behairy NH et al. Egypt J Radiol Nucl Med (2017), https://doi.org/10.1016/j.ejrnm.2017.07.002