The quadricuspid aortic valve

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ABSTRACT

The quadricuspid aortic valve is a very rare congenital defect that may be an isolated entity or associated with a multitude of cardiovascular abnormalities. Isolated cases usually manifest late in life with the complication of aortic regurgitation, which can be delayed by early valve replacement. We present a case of quadricuspid aortic valve associated with truncus arteriosus, ventricular septal defect, and interrupted aortic arch.

CASE REPORT

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A 5-year-old girl was seen in the cardiac outpatient clinic. The patient was born prematurely, at an estimated gestational age of 31-35 weeks. The history revealed status post truncus arteriosus repair and interrupted aortic arch repair with ventricular septal defect patch closure, aortic arch augmentation with pulmonary homograft patch, and right ventricular outflow tract to the main pulmonary artery conduit. A cardiac magnetic resonance imaging (MRI) was performed to evaluate the patency of the conduit (fig. 1), which revealed a regurgitant jet across the aortic valve. Incidentally, a quadricupid aortic valve (Fig. 2) was noted. Software based analysis (Argus; Siemens Medical Systems) demonstrated a regurgitant fraction of 32% (Fig. 3, 4), which is categorized as a moderate severity of aortic regurgitation. The patient is scheduled for an annual follow-up by cardiac MRI. Based on the severity of the aortic valve regurgitation, the size of the conduit, and the clinical presentation, a decision regarding the time for aortic valve replacement will be made.

DISCUSSION

Etiology and demographics

A quadricuspid aortic valve was first identified on autopsy by Balington in 1862 [1]; the first in vivo description dates back to 1968 by Robicsek et al [1]. A quadricuspid aortic valve usually appears as an isolated congenital anomaly, but may also be associated with other malformations, the most common being coronary artery anomalies [2]. The quadricuspid aortic valve is a rare manifestation with an incidence of 1% in a review of patients undergoing surgery for isolated aortic regurgitation [3, 4]. In the general populations the prevalence of a quadricuspid aortic valve was reported to be in the range of 0.013-0.043% [5, 6].

Clinical & Imaging Findings

Several different anatomical variations of a quadricuspid aortic valve have been described as follows: Type A - four equal cusps; type B - three equal cusps and one smaller cusp; type C - two equal larger cusps and two equal smaller cusps; type D - one large, two intermediate and one small cusp; type E - three equal cusps and one larger cusp; type F - two equal larger cusps and two unequal smaller cusps; and type G - four unequal cusps [7, 8]. Types A and B are the most common [5]. A quadricuspid aortic valve is rarely diagnosed in children, but it can be associated with significant aortic regurgitation [9]. Patients tend to present clinical symptoms in their 5th or 6th decade of life. The physiopathology of the valve dysfunction is poorly understood: anatomical abnormalities of the cusps could induce unequal shear stress leading to fibrosis and incomplete coaptation [5]. As in bicuspid valve, regurgitation of an abnormal aortic valve may also occur as a result of a prolapse of the layer of the cusps or be associated with aortic root dilatation [3, 10].

The literature reports the association of a quadricuspid aortic valve with various congenital defects such as partial anomalous pulmonary venous return with atrial septal defect [11], tetralogy of Fallot [12], patent ductus arteriosus [13],

anomalous origin of the coronary arteries [14] or truncal anomalies [15], as in the case described here.

The diagnosis of a quadricuspid valve is often confirmed by imaging such as computerized tomography (CT), MRI, or echocardiography. Most of the time when associated with other congenital anomalies, a quadricuspid valve is diagnosed incidentally during infancy or childhood. When the entity is isolated, the diagnosis is made later in combination with the assessment for aortic regurgitation.

Differential Diagnosis

Although the diagnosis of a quadriscupid valve is typically straight forward based on its typical morphological features, there may be a differential diagnosis, e.g. in cases when the valvular anatomy is not clearly delineated for a variety of reasons. The differential diagnosis may include neoplastic involvement of the valve, valvular degeneration with or without calcifications, adherent thrombus or vegetations. Among the tumors associated with the aortic valve are papillary fibroelastoma, which is the most common valvular tumor, followed by myxoma. Cine gradient-echo MRI is typically able to depict even small tumors attached to moving cardiac valve leaflets. An organized thrombus attached to the valve may be isolated or associated with other disease entities and may occasionally resemble an additional cusp. Calcifications and vegetation with or without endocarditis are another differential consideration. At times cusps may be fused, which may give the appearance of a bicuspid valve. Diagnosis should rely on the combination of clinical presentation and morphology of the valve as determined by imaging.

Treatment & Prognosis

The mainstay of treatment is aortic valve replacement [16]. The treatment can be associated with aortic root graft replacement, if aneurysmal dilatation of the aortic root has occurred. In patients with additional congenital cardiac anomalies, the aortic valve is usually repaired at an earlier stage. For an uncomplicated case aortic valve replacement may be performed through a transaortic approach. The prognosis is excellent if the case is isolated, without associated congenital abnormalities, and if the valve was replaced prior to the occurrence of complications.

Conclusion

Most of the imaging studies in the literature are based on echocardiography, as this modality is most widely used in this context. However, with CT and MRI as emerging modalities for cardiac imaging, it is important to recognize this abnormality in cross sectional imaging [17].

TEACHING POINT

As cardiac CT and MRI are becoming more widely available, it is essential that this rare condition (quadricuspid aortic valve) is recognized. Patents with an isolated quadricuspid valve may clinically present later in life. In patients with a quadricuspid aortic valve follow-up evaluations are important in order to enable early treatment and therefore to prevent complications.

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FIGURES

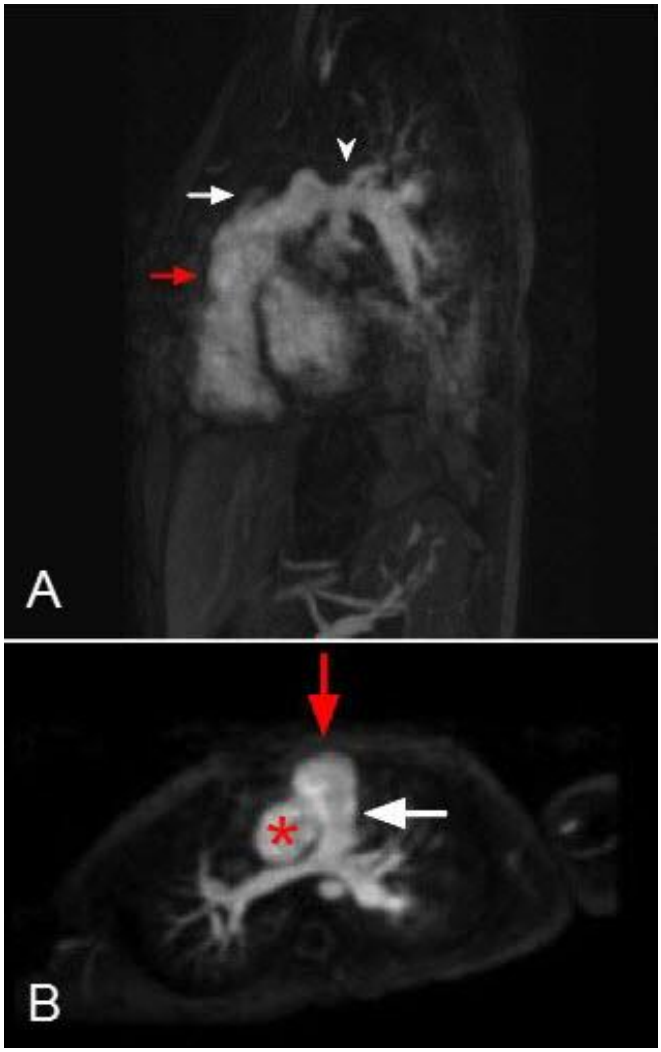


Figure 1 (left): 5-year-old female with truncus arteriosus repair and quadricuspid aortic valve. Oblique sagittal (A) and axial (B) thin maximum intensity projection (MIP) MR images obtained on a Siemens TimTrio 3.0T scanner using a 3D FLASH sequence (TR/TE/flip angle: 2.8ms/1.06ms/18°; matrix: 292 x 576; FoV: 281 x 500) following the intravenous administration of a single dose (5 mL) of Gd-based paramagnetic contrast agent (gadobenate dimeglumine), demonstrates a widely patent conduit (white arrow) between the RVOT (red arrow) and the main pulmonary artery (white arrowhead). (Red asterisk - aorta)

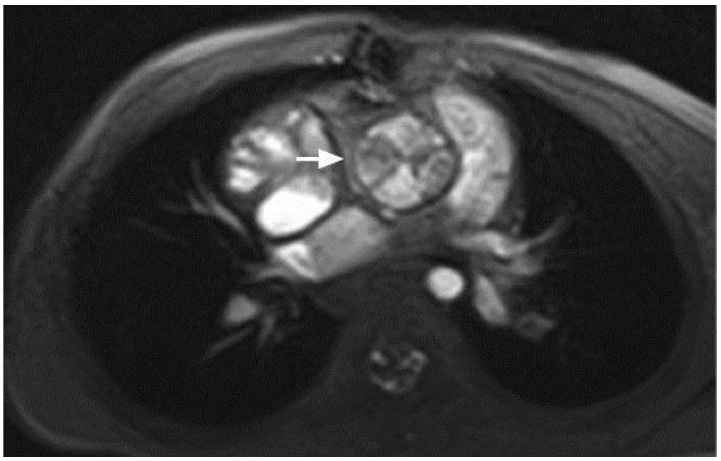
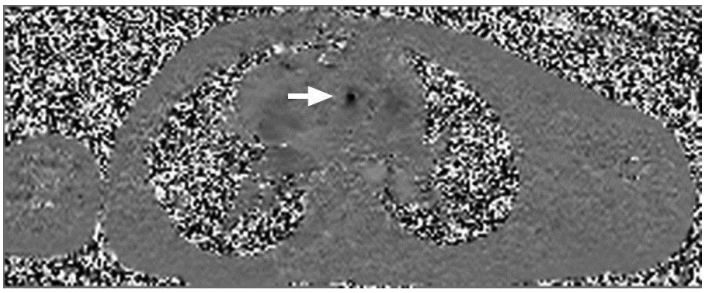


Figure 2: 5-year-old female post truncus arteriosus repair with type A quadricuspid aortic valve. Axial image obtained on a Siemens TimTrio 3.0T scanner using a dynamic cine 2D spoiled gradient echo sequence (FLASH; TR/TE/flip angle: 38.07/1.88/15°; matrix: 292 x 576; FoV: 131 x 300mm) demonstrates a quadricuspid aortic valve (arrow); the four isolated aortic leaflets are clearly depicted.

Figure 3 (right): 5-year-old female post truncus arteriosus repair and quadricuspid aortic valve. Axial phase encoded image of a phase contrast sequence (fl2D; TR/TE/flip angle: 39.5/2.17/30°; matrix: 94 x 256, FoV: 121 x 300mm) obtained on a Siemens TimTrio 3.0T scanner demonstrates flow reversal (regurgitation) across aortic valve indicated by dark signal (arrow) due to incomplete coaptation of aortic valve cusps.



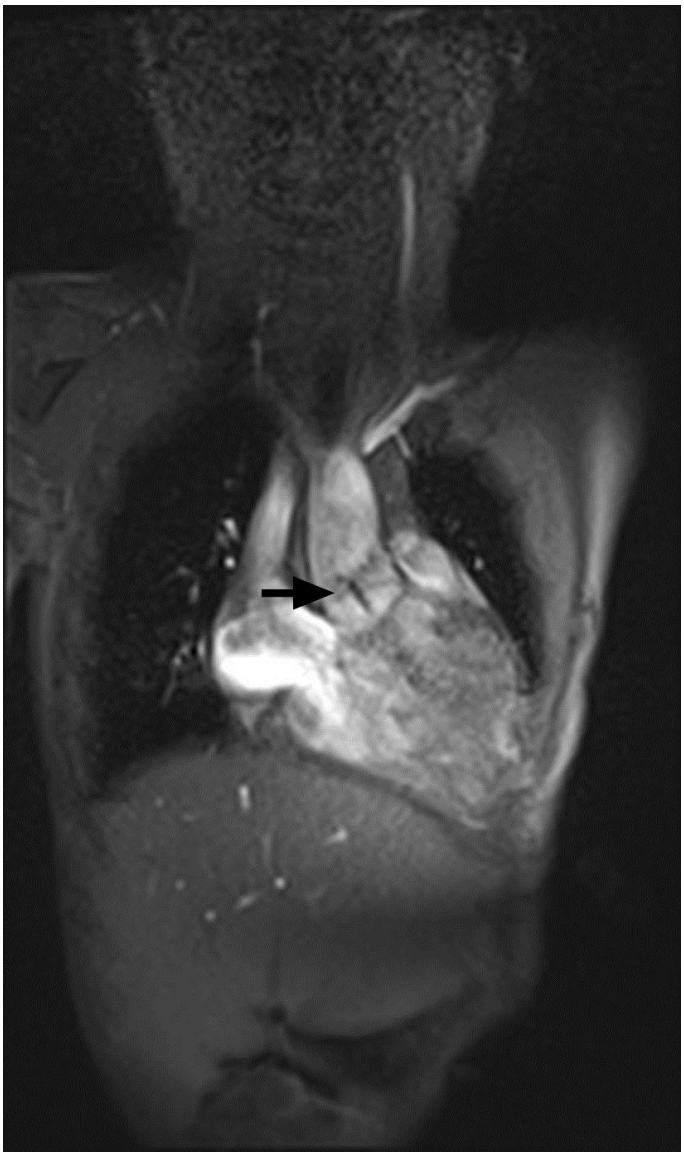


Figure 4: 5-year-old female post truncus arteriosus repair and quadricuspid aortic valve. Right ventricular outflow tract (RVOT) projection image of cardiac MRI obtained on a Siemens TimTrio 3.0T scanner using a cine FLASH sequence (TR/TE/flip angle: 36.8ms/1.8ms/0°) demonstrates regurgitant jet across the aortic valve into the left ventricular outflow tract (arrow), indicating aortic valve regurgitation (32% regurgitant fraction per software based quantitative analysis).

Etiology	Congenital
Incidence	0.013–0.043%
Gender ratio	1:1
Age predilection	With an isolated case without associated findings, clinical manifestations tend to appear between the 5th and the 6th decades of life.
Risk factor	Unknown
Treatment	Transaortic or surgical aortic valve replacement. In case of complications aortic root is grafted.
Prognosis	Excellent, if no associated congenital abnormalities
Findings on imaging	Four aortic cusps visualized in cross sectional imaging

Table 1: Summary table for quadricuspid aortic valve

Disease	**MRI**	**CT**
Papillary fibroelastoma	Low signal in T2, identical to fibrous tissue.	Regular or irregular contours of the tumor with no enhancement
Myxoma	Low signal in T1 and heterogeneous signal in T2. Enhancement can distinguish from thrombus	Heterogeneously low signal due to calcifications or hemorrhage
Thrombus	Low signal in T1 and T2 without enhancement	Low attenuation filling defect
Calcifications	Low signal thickening of the valve	High attenuating foci within the cusps.
Vegetation	Low T1; heterogeneous signal in T2. Enhancement if infected	Filling defect that enhances if infected
Bicuspid aortic valve	Two cusps with fish-mouth opening in cine views	Two cusps with fish-mouth opening in cine views

Table 2: Differential diagnosis table for quadricuspid aortic valve when the valvular anatomy is not clearly delineated

ABBREVIATIONS

CT - Computerized tomography
FLASH - Fast Low Angle Shot
HASTE - Half-Fourier acquisition single-shot turbo spin-echo
LVOT - Left ventricular outflow tract
MIP - Maximal Intensity Projection
MRI - Magnetic resonance imaging
RVOT - Right ventricular outflow tract

KEYWORDS

Quadricuspid aortic valve; Heart; Truncus Arteriosus; Cardiac valves; cardiac Magnetic resonance Imaging

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