# A rare pediatric cardiac anomaly: Quadricuspid aortic valve with aortic regurgitation

**Michael Brock, Aykut Tugertimur[1], Matthew C Schwartz[1]**
Department of Pediatrics, Arnold Palmer Hospital for Children, [1]The Heart Center, Arnold Palmer Hospital for Children, Orlando, Florida, USA

## ABSTRACT

**A quadricuspid aortic valve is rarely diagnosed in children, but it can be associated with significant aortic regurgitation. It is important for pediatric cardiologists to be aware of this pathologic entity. We present a nine-year-old male, diagnosed with a quadricuspid aortic valve and mild aortic regurgitation.**

**Keywords:** Aortic valve, aortic regurgitation, quadricuspid

## CASE REPORT

A 9-year-old male was referred to the cardiology clinic for evaluation of an asymptomatic diastolic heart murmur. On physical examination, a 2/4 early, medium-frequency, diastolic murmur was audible at the right mid-sternal border. The remainder of his physical examination was unremarkable and an electrocardiogram was normal. Transthoracic echocardiography showed a type B quadricuspid aortic valve (QAV) (three cusps of equal size and one smaller cusp) associated with mild aortic regurgitation by color Doppler imaging, likely from a coaptation defect [Figure 1 and Video 1].[1] The left ventricular end-diastolic dimension measured by M-mode was normal (4.58 cm, z-score + 0.3) with a shortening fraction of 31%. The remainder of the cardiac anatomy was normal, including the origins of the coronary arteries.

Two years later, the patient remained asymptomatic and the echocardiography showed stable, mild aortic regurgitation [Figure 2], now associated with mild left ventricular dilation [Figure 3]. By M-mode the left ventricular end-diastolic dimension measured 5.82 cm (z-score + 2.8), the end-systolic dimension was 3.7 cm (z-score + 1.9), and the shortening fraction was 35%.

Video available on www.annalspc.com

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/0974-2069.115285

## DISCUSSION

A QAV is an extremely rare congenital heart malformation. Often found incidentally during an autopsy, the incidence is between 0.008 and 0.043%.[2,3] Diagnosis of a QAV during childhood is uncommon, as most are diagnosed in adulthood. The mean age of diagnosis is 50.7 years and there is a slight male predominance.[4]

Aortic regurgitation is the most prevalent hemodynamic abnormality associated with a QAV and appears in up to 75% of the patients.[4] This rarely develops in childhood and is thought to be a consequence of mechanical stress leading

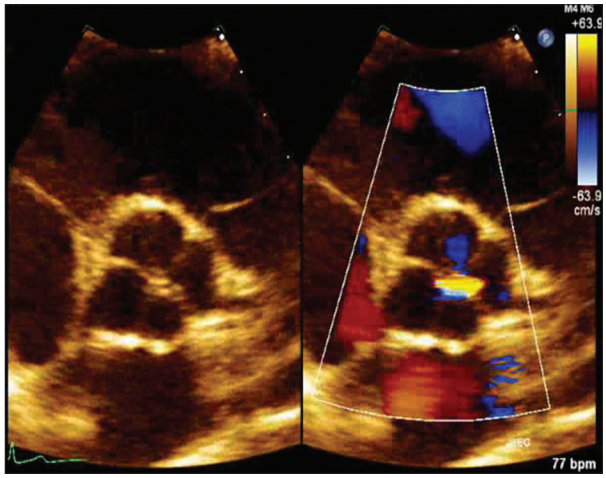


**Figure 1: Parasternal short axis view of the transthoracic echocardiogram showing two-dimensional and color Doppler diastolic images of a quadricuspid aortic valve with mild insufficiency, due to abnormal leaflet coaptation, in a nine-year-old male**

**Address for correspondence:** Dr. Michael Brock, Arnold Palmer Hospital for Children, 92 West Miller Street, Orlando, Florida 32806.
E-mail: michael.brock@orlandohealth.com

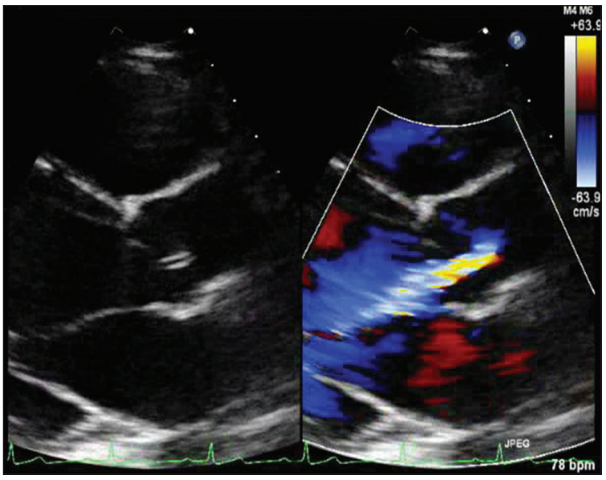


**Figure 2: Parasternal long axis view of the transthoracic echocardiogram showing two-dimensional and color Doppler diastolic images in the same patient shown in Figure 1, now 11-years-old. The patient has mild aortic insufficiency due to malcoaptation of the quadricuspid aortic valve leaflets**

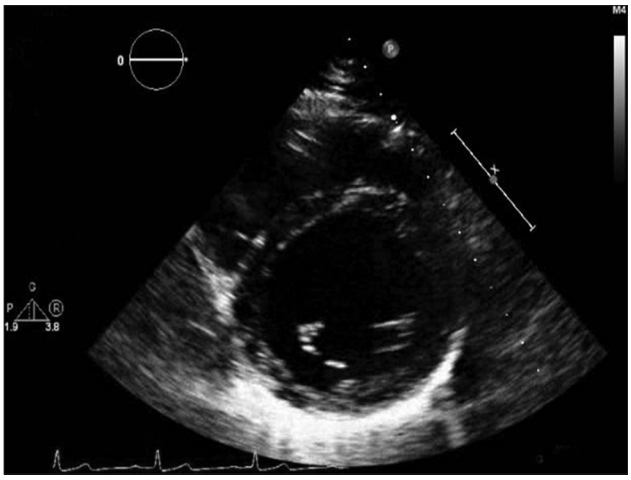

**Figure 3: Parasternal short axis view of the transthoracic echocardiogram showing mild left ventricular dilation in the 11-year-old male**

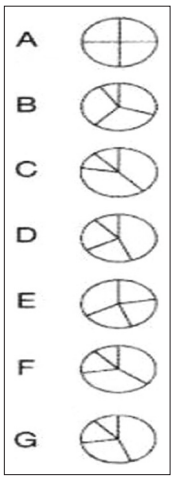


**Figure 4: Seven anatomic variations of quadricuspid aortic valves, as described by the authors of reference 1. The figure has been adapted from its original presentation in reference 1, with permission. Type A shows four equal cusps; type B shows three equal cusps and one smaller cusp; type C shows two equal larger cusps and two equal smaller cusps; type D shows one large, two intermediate, and one small cusp; type E shows three equal cusps and one larger cusp; type F shows two equal larger cusps and two unequal smaller cusps; type G shows four unequal cusps**

to leaflet fibrosis and malcoaptation over time.[1] Many require aortic valve replacement later in adulthood.[4] Our patient exhibited mild aortic insufficiency due to abnormal leaflet coaptation at only nine years of age, which caused a diastolic murmur and led to his early diagnosis.

Unlike bicuspid aortic valve, aortic stenosis is rare among those with a QAV.[4] Up to 18% of the patients will have other cardiac malformations, most common being a coronary artery anomaly.[4] Atrial septal defect, pulmonary valve stenosis, hypertrophic cardiomyopathy, ventricular septal defect, and subaortic stenosis have also been described.[4,5]

Hurwitz and Roberts classified QAV into seven types, A through G [Figure 4].[1] Our patient had a type B valve with three cusps of equal size and one smaller cusp. Types A and B are the most common forms.[3,5] Some have argued that type B valves are more likely to develop regurgitation because the single smaller cusp leads to unequal stress distribution, progressive trauma, and malcoaptation, with age.[3] However, a recent review suggests that both forms are often regurgitant.[5]

Although QAV is rare and uncommonly diagnosed in children, pediatric cardiologists should be familiar with this congenital malformation and its associated cardiac anomalies. Currently, our patient has mild aortic regurgitation with mild left ventricular dilation and preserved shortening. He will require a lifelong cardiology follow-up, with a possible need for intervention later in life.

## REFERENCES

1. Hurwitz LE, Roberts WC. Quadricuspid semilunar valve. Am J Cardiol 1973;31:623-6.
2. Simonds JP. Congenital malformation of the aortic and pulmonary valves. Am J Med Sci 1923;166:584-95.
3. Feldman BJ, Khanderia BK, Warnes CA, Sweard JB, Taylor CL, Tajik AJ. Incidence, description and functional assessment of isolated quadricuspid aortic valves. Am J Cardiol 1990;65:937-8.
4. Tutarel O. The quadricuspid aortic valve: A comprehensive review. J Heart Valve Dis 2004;13:534-7.
5. Timperley J, Milner R, Marshall AJ, Gilbert TJ. Quadricuspid aortic valves. Clin Cardiol 2002;25:548-52.

**How to cite this article:** Brock M, Tugertimur A, Schwartz MC. A rare pediatric cardiac anomaly: Quadricuspid aortic valve with aortic regurgitation. Ann Pediatr Card 2013;6:202-3.

**Source of Support:** Nil, **Conflict of Interest:** None declared