Heart Defect Screening Indicated in Turner's

BY MIRIAM E. TUCKER

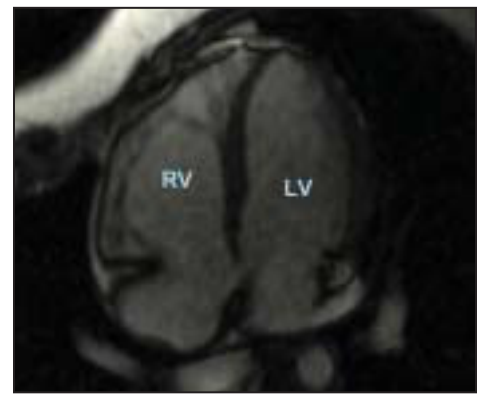
NEW YORK — Partial anomalous pulmonary venous return was detected by cardiac magnetic resonance imaging in 7 of 39 adolescent and young adult women with Turner's syndrome whose charts were retrospectively evaluated.

The finding suggests that screening for the partial anomalous pulmonary venous return (PAPVR) heart defect is indicated in Turner's syndrome patients, along with appropriate cardiac referral and management, Dr. Iris Gutmark-Little said at the joint meeting of the Lawson Wilkins Pediatric Endocrine Society/European Society for Pediatric Endocrinology.

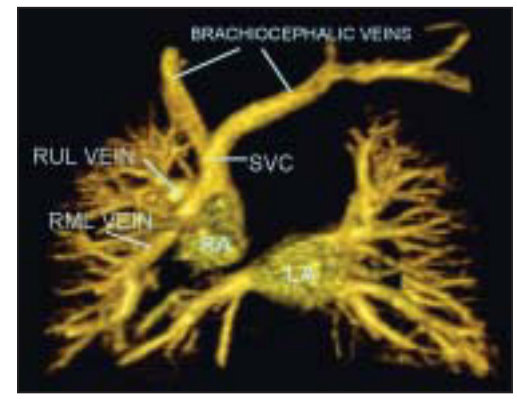
The results also suggest that cardiac magnetic resonance (CMR) imaging is more sensitive than echocardiography for the detection of PAPVR in Turner's patients, said Dr. Gutmark-Little of Cincinnati Children's Hospital Medical Center.

The 39 patients were the first to be screened with CMR after Cincinnati Children's began using the modality routinely in all Turner's patients during their early teen years. The patients also underwent echocardiographic evaluation.

Seven patients (18%) were found to have PAPVR by cardiac magnetic resonance imaging, six of them following a normal echocardiogram. Aberrant drainage of the right upper pulmonary veins was seen in five patients, three of whom had involvement of at least a portion of the right middle lobe vein. The other two had the defect in the left upper pulmonary vein.



A four-chamber MRI view shows a dilated right ventricle resulting from a right-sided PAPVR lesion in a Turner's patient.



Three-dimensional MRI of a right-sided PAPVR lesion shows anomalous pulmonary veins draining into the superior vena cava.

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The 18% PAPVR prevalence seen here is similar to the 13% found in a previous study of adult Turner's patients (Circulation 2004;110:1694-700).

In one patient, PAPVR was associated with clinically significant enlargement of the right ventricle, with a pulmonary-to-systemic blood flow ratio (Qp:Qs) of 1.9:1. She required surgical repair. The other six patients had Qp:Qs ratios ranging from 1.24:1 to 1.62:1 and did not require intervention.

There were no differences in age, height, karyotype, or right ventricular ejection fraction between the 7 patients with PAPVR and the 32 without, nor did the two groups differ in peripheral lymphedema, neck webbing, renal malformations, coarctation, or bicuspid aortic valve. This finding differs from previous studies that have linked some of these features to PAPVR in Turner's patients, said Dr. Gutmark-Little, who stated that she had no financial disclosures. ■

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