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Research article

Partial anomalous pulmonary venous return in Turner syndrome

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ABSTRACT

Purpose: The aim of this study is to describe the prevalence, anatomy, associations and clinical impact of partial anomalous pulmonary venous return in patients with Turner syndrome.

Methods and results: All Turner patients who presented at our Turner clinic, between January 2007 and October 2015 were included in this study and underwent ECG, echocardiography and advanced imaging such as cardiac magnetic resonance or computed tomography as part of their regular clinical workup. All imaging was re-evaluated and detailed anatomy was described. Partial anomalous pulmonary venous return was diagnosed in 24 (25%) out of 96 Turner patients included and 14 (58%) of these 24 partial anomalous pulmonary venous return had not been reported previously. Right atrial or ventricular dilatation was present in 11 (46%) of 24 partial anomalous pulmonary venous return patients.

Conclusion: When studied with advanced imaging modalities and looked for with specific attention, PAPVR is found in 1 out of 4 Turner patients. Half of these patients had right atrial and/or ventricular dilatation. Evaluation of pulmonary venous return should be included in the standard protocol in all Turner patients.

1. Introduction

Turner syndrome (TS) is a partial or complete monosomy of the X-chromosome, and was originally described by Henry Turner in 1938. It occurs in 1 per 2500 live born females [1–3]. Turner syndrome is nowadays often diagnosed prenatally or during early childhood and patients present with a broad variety of disorders, including short stature, estrogen deficiency and cardiovascular abnormalities [4]. Aside from a higher risk profile for ischemic heart disease, often congenital cardiac defects, located mainly on the left side, can be found in these patients. The most common lesions are: elongation of the aortic arch (49%), bicuspid aortic valve (14–30%), coarctation (7–18%), persistent left superior vena cava (13%), and atrial and ventricular septal defect (0–8%) [2,5–9]. PAPVR has been described to be more prevalent in TS patients, however knowledge about detailed anatomy and precise prevalence is scarce, partly because it is not routinely looked for at first recognition of the syndrome [6–9]. However, PAPVR can cause an hemodynamically significant left-to-right shunt and subsequently, right chamber dilatation, arrhythmias and even pulmonary hypertension [10]. This necessitates early diagnosis and, in case of a large shunt, treatment. In our tertiary center for Turner syndrome all adult patients undergo a CT scan, while children and adolescents will have an MRI [11].

The aim of this study is to describe the prevalence, anatomy and clinical significance of PAPVR, in patients with Turner syndrome. Furthermore, factors, possibly related with PAPVR were studied.

2. Methods

Every Turner patient is enrolled into a prospective surveillance program according to a local standardized clinical protocol. This protocol includes clinical assessment by a cardiologist, ECG,

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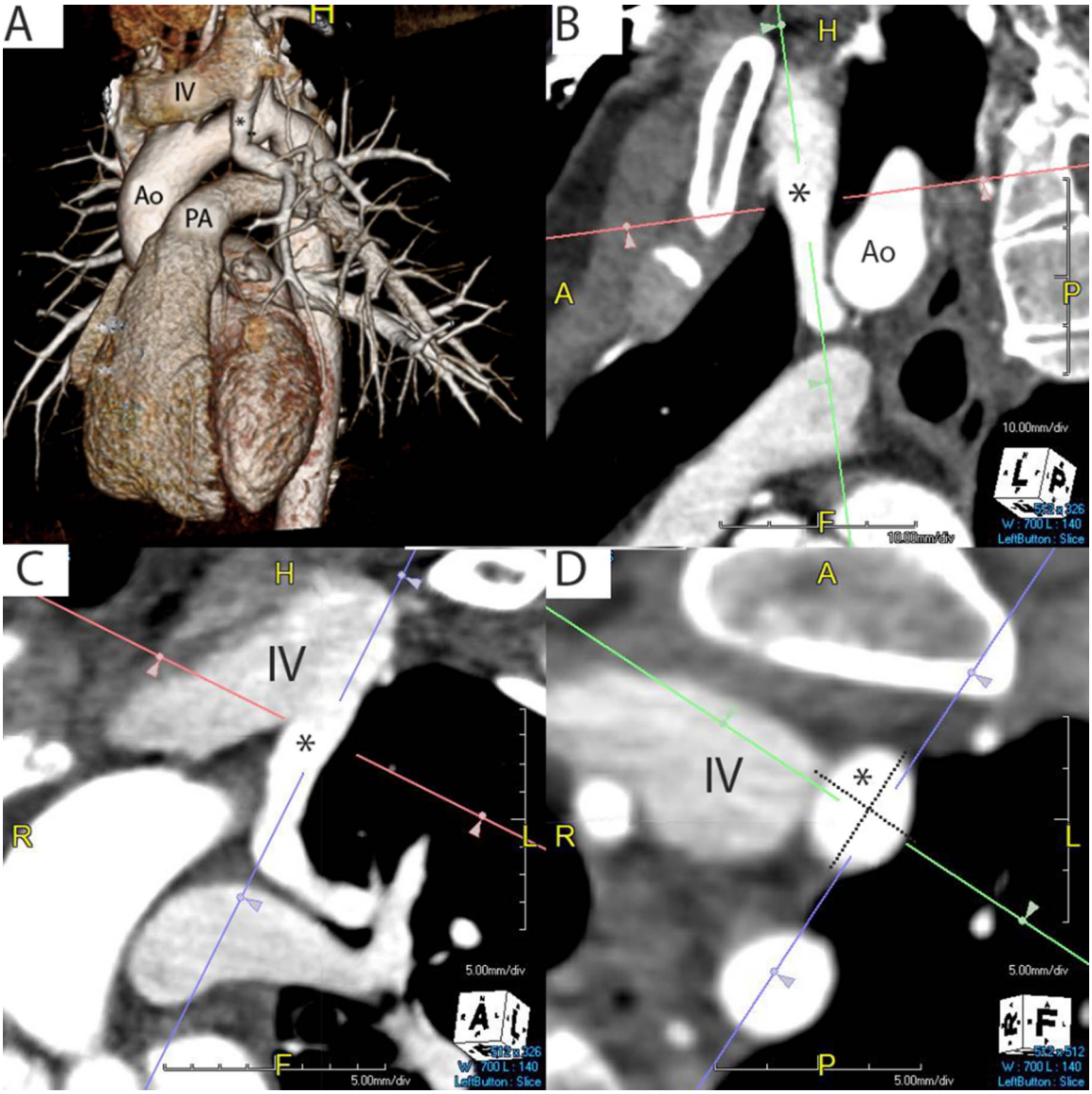

Fig. 1. Measurement of PAPVR. Panel A) 3D volume rendered reconstruction showing a PAPVR (*), the Aorta (Ao), Pulmonary Artery (PA), innominate vein (IV). Panel B and C) Sagittal and coronal oblique views of how the connecting site area of the PAPVR (*) was measured. D) Cross section of the PAPVR (*) perpendicular to the vessel.

echocardiography and cardiac CT (adults) or MR imaging (children and adolescents). For this study all adult patients, who were evaluated at the department of congenital cardiology between January 2007 and October 2015 with genetically proven Turner syndrome were included. We excluded patients with insufficient CT or MR imaging quality. The study was approved by the medical ethical committee of our center. Informed consent was waived.

2.1. Imaging

For this study all CT and MR-images were re-evaluated by two independent investigators (A.H. and R.C.), blinded to each other. When more than one scan (CT or MR) was available per patient, the most recent CT scan was used. Evaluation was done by multi planar reconstruction, using the AquariusNet (TeraRecon, Inc, San Mateo, CA) software. The connecting site was defined as the location where the anomalous pulmonary vein drained into the systemic venous system. At this location vessel diameter and area were measured; perpendicular to the anomalous vein, as proximal to the vessel it feeds into as possible but where it was still discernable as a separate vessel (Fig. 1.) The anomalous vessel was then traced back to the lung lobe or lobes from which it originated; the site of origin. Right atrial and right ventricular dilatation was visually assessed on echocardiography images by two observers (A.T. and J.R.), blinded to CT/MR data, on apical 4-chamber view, parasternal long-axis and parasternal short-axis according to the latest guidelines [12] (Fig. 2). To assess global RV systolic function the right ventricular fractional area change (RV FAC) was calculated, where a RV FAC < 32% was considered impaired [12].

2.2. Computer tomography angiography (CTA)

For this retrospective study, most patients (n = 32, 33%) were scanned according to a now standard protocol where a retrospectively ECG-triggered CTA was obtained on a dual source scanner (Force or Flash, Siemens, Germany). Sixty-five ml of iodinated contrast material (Iodixanol, GE, USA) was administered through an antecubital vein followed by a 40 ml 70/30% saline/contrast chaser, both at 5 ml/second flow rates. Images were reconstructed at each 2% for 0–40% of the R–R interval with 1.0 mm/0.6 mm thickness and interval. Also, images at each 5% of the entire R–R interval were reconstructed at 1.5 mm with a 1.0 mm interval. Maximal aortic diameters in CTA were generated using the double-oblique short-axis technique. Another 32