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Importance of carefully interpreting computed tomography images to detect partial anomalous pulmonary venous return

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ABSTRACT

Background: Partial anomalous pulmonary venous return (PAPVR) is characterized by an abnormal connection of the pulmonary vein (PV). The left-to-right shunt results in an increased pulmonary blood flow, which may be followed by developing pulmonary hypertension (PH). We found that computed tomography (CT) scans may be misinterpreted, potentially leaving anomalous PVs undetected when reviewing diagnostic findings of PAPVR patients. The purpose of this study was to delineate this risk and assess the usefulness of our interpretation methods.

Methods: We retrospectively reviewed the records of 8 patients diagnosed with PAPVR, diagnosed with right heart catheterization (RHC) findings, at our department between 1991 and 2013. Our CT screening method for assessing anomalous PVs consisted of two points: 1) confirming that four PVs were connected to the left atrium (LA) and 2) checking that the vena cava was not connected with anomalous PVs. The accuracy of this method was analyzed in a blinded manner.

Results: In 4 patients, anomalous PVs delineated on enhanced CT scan images obtained before RHC were undetected. The sensitivity and specificity of detecting PAPVRs using our protocol were 0.800 and 0.978, respectively. Four of 8 patients went on to develop PH. Age at the time of diagnosis was positively correlated with mean pulmonary arterial pressure ($r=0.929$, $p=0.002$).

Conclusion: There is a potential risk of CT scan misinterpretation when looking for anomalous PVs. Careful interpretation of CT findings that focus on PVs may be useful for detecting PAPVR and obtaining a PH differential diagnosis.

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1. Introduction

Pulmonary anomalous pulmonary venous return (PAPVR) is an uncommon congenital heart defect characterized by abnormal connections of up to three pulmonary veins (PVs), mainly to the vena cava and/or right atrium [1]. It was previously reported that PAPVR is found in 0.4–0.7% of Caucasian autopsy cases [2–4] and in 0.05% of elderly Japanese patients [5]. Pulmonary veins drain into systemic veins (left-to-right shunt), and, in patients with this disease, shunted blood travels through the pulmonary circulation again [1]. This increase in pulmonary blood flow may result in the onset of pulmonary hypertension (PH) and right heart failure [3,4]. We diagnosed PAPVR in 8 of 976 patients (0.8%) undergoing right heart catheterization (RHC) at our institution between 1991 and 2013. Computed tomography (CT) scans were commonly performed to determine whether patients had PH associated with chronic lung disease or chronic thromboembolic pulmonary hypertension (CTEPH) [6]. We found that CT scans may be misinterpreted when clinical characteristics and diagnostic work-up findings of PAPVR patients are reviewed. This may result in anomalous PVs remaining undetected. The purpose of this study was to analyze the potential risk of not detecting anomalous PVs and to determine the usefulness of CT scan interpretation methods.

2. Patients and methods

2.1. Ethical statement

According to Japanese legislation, informed consent is not required for the retrospective collection of data that relate to current practice. However, the study database was anonymized and complied with the requirements of the Ministry of Health, Labour and Welfare, which is dedicated to privacy, information technology, and civil rights in Japan. The Ethics Committee of Chiba University approved the study protocol (Approval date: 27 Oct, 2011. approval number: 1248).

2.2. Patient characteristics

Eight of 976 patients (0.8%) who underwent RHC at the Chiba University Hospital respiratory ward between 1991 and 2013 were diagnosed with PAPVR. Data for these patients were retrospectively reviewed. Case 5 has been reported previously [7]. All patients who developed PH during their clinical course were considered to have PAPVR with PH.

2.3. Computed tomography scans

Seven of 8 PAPVR patients (87.5%) had CT scans performed before diagnostic RHC and pulmonary angiography (PAG). In Case 1, the patient did not undergo CT scanning before RHC and PAG because, at the time of diagnosis, multidetector CT had not yet been introduced at our hospital. In Case 6, the patient underwent two CT scans before RHC because the initial evaluation had been performed at another hospital. Details of instruments and settings are provided in Supplemental Table 1. Contrast material (100 ml of 350 mg/ml of iodine) was intravenously injected at a rate of 3 ml/s using a mechanical injector. All CT examinations were performed with a scanning delay of 20–30 s for optimal pulmonary artery visualization.

2.4. Computed tomography scan interpretation and pulmonary anomalous pulmonary venous return diagnosis

The protocol for interpreting CT scans is shown in Fig. 1. Anomalous PVs were screened using CT scans using the following criteria: 1) presence of four PVs connected to the left atrium (LA) and 2) absence of anomalous PV connections to the vena cava. If either or both of these criteria were not met, the patient was suspected to have PAPVR. To determine detection accuracy, CT scan images of 50 patients (5 PAPVR patients [all right-sided]), 45 control patients) were examined by an investigator (TS). Two trained pulmonologists (TJ, AN) that were blinded to patient status independently interpreted CT scan images using the above protocol.

2.5. Right heart catheterization and pulmonary angiography

A 7.5 Fr Swan-Ganz catheter (Edwards Lifescience, Irvine, CA, USA) was inserted from the right or left internal jugular vein. Blood pressure and gas values were measured at the following sites: superior vena cava (SVC), inferior vena cava (IVC), right atrium (RA), right ventricle (RV), main pulmonary artery, and right or left pulmonary artery. The pulmonary arterial wedge pressure (PAWP) was also measured, along with PV and LA pressure via an atrial septal defect (ASD) if present. Cardiac output and cardiac index calculations were based on the Fick principle. The pulmonary to systemic blood flow (Qp/Qs) ratio was calculated using these data. Pulmonary angiography was performed at the time of RHC. Iodinated contrast material (18–20 ml) was injected through a 7 Fr Berman catheter at a flow rate of 9–10 ml/s. Angiogram images were obtained at a rate of 2–3 frames/s.

2.6. Ultrasonic cardiograms

Transthoracic echocardiography (TTE) was performed by trained cardiologists at our institution. The systolic transtricuspid pressure gradient (TRPG) was calculated using the Bernoulli equation, which states:

$$TRPG = 4 \times v^2$$

where v is the maximum velocity of blood through the tricuspid (m/s). The RA pressure was estimated based on the method used by Kircher [8]. Estimated systolic PA

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