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Diastolic Pressure Alternans: A New Sign in Congenital Absence of the Pericardium

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ABSTRACT

Congenital absence of the pericardium is a rare condition, which is frequently missed due its subtle presentation. It may be misdiagnosed as another condition causing right heart dilatation such as an intracardiac shunt. We report the finding of diastolic pressure alternans during cardiac catheterization in this single case report of a patient with congenital total absence of the pericardium. The occurrence of this phenomenon is hypothesized to be due to excessive cardiac hypermobility and paradoxical septal movement. We propose that this finding may be a useful clue to the diagnosis.

Keywords: Congenital absence of the pericardium, cardiac catheterization, diastolic pressure alternans

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CASE REPORT

A 37-year-old Malay gentleman was admitted to the surgical ward for an elective laparotomy for a carcinoma of the pancreas. He was asymptomatic cardiac-wise but was noted to have an abnormal chest X-ray (Fig. 1) pre-operatively and referred to the attending cardiologist for an opinion. Physical examination revealed that the apex beat was prominent and was in the 4th intercostal space in the anterior axillary line. On auscultation, the second heart sound was widely split but no murmurs were audible. The ECG showed right axis deviation, a tall R wave in V1 and V2 with late QRS transition. The diagnosis of congenital absence of the pericardium was not considered at this point (although retrospectively the physical examination and chest X-ray showed the classical features of the condition) but he was listed for an echocardiographic examination for a suspected atrial septal defect. The echo revealed an enlarged right ventricle and right atrium, but no intracardiac shunts could be demonstrated and an echo window in the 3rd intercostal space in the midclavicular line had to be used to obtain the conventional parasternal long and short axis views. Congenital absence of the pericardium was then suspected, but as the echocardiographic window was not entirely satisfactory and there was a need for a definitive diagnosis before major abdominal surgery, cardiac catheterization was performed. Left and right heart catheterisation and oximetry revealed normal pulmonary pressures and no intracardiac shunts. During simultaneous right and left ventricular pressure measurements, it was noticed that the patient exhibited regular alternans of the diastolic pressures

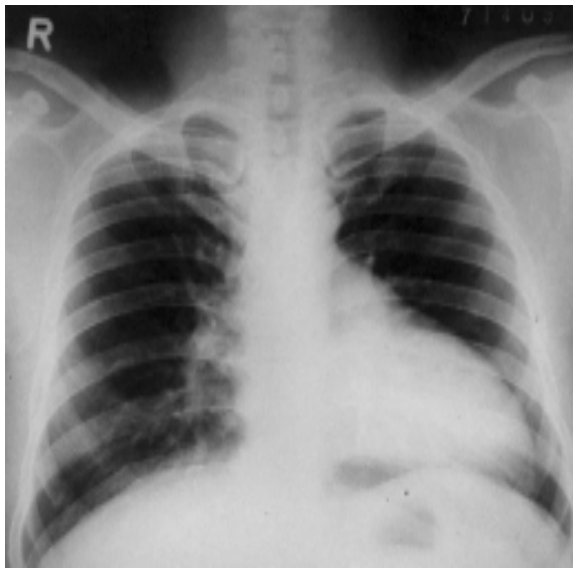

Fig. 1 Postero-anterior chest X-ray of the patient showing the enlarged heart silhouette and the interposition of the lung between the inferior surface of the heart and the diaphragm.

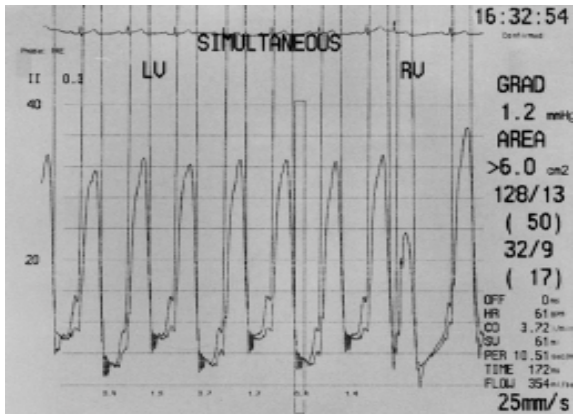

Fig. 2 Pressure tracing of the left and right ventricle showing diastolic pressure alternans.

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(Fig. 2). The diastolic pressures in the right and left ventricle were also equal with overlapping tracings. An echocardiogram was then performed on the catheterization table and this revealed that there was paradoxical septal motion with excessive cardiac hypermobility, features that were noted during the previous examination. The beat-to-beat, to-and-fro motion of the heart corresponded to the regular variation of the ventricular diastolic pressures. Left and right ventriculography was also performed and this too showed the excessive to-and-fro swinging of both ventricles with each cardiac cycle. A CT scan, which was subsequently ordered after the catheterization study, confirmed the absence of the left pericardium.

DISCUSSION

Congenital total absence of the pericardium is a condition that is frequently missed antemortem and pre-operatively because the patients are usually asymptomatic and the signs due to the lack of pericardial restraint (hypermobility) are subtle. The patients who have a partial pericardial defect do not have features of hypermobility but may complain of symptoms of chest pain or may have syncope or sudden death due to incarceration of cardiac tissue[1] and both varieties can be associated with severe tricuspid regurgitation secondary to chordal rupture[2,3]. Patients have been subjected to cardiac catheterization with an echo diagnosis of right heart dilatation secondary to an intracardiac shunt such as an atrial septal defect only to find no defect during catheterisation[4]. ln our patient, we observed the interesting finding of diastolic pressure alternans.

It is known that the ventricular volumes in these patients are subject to much variation due to the absence of pericardial restraint[5]. Diastolic pressure alternans is presumed to occur because there is excessive hypermobility with a beat-to-beat, to-and-fro swinging of the heart and distortion of the ventricular shape and volume during each cycle coupled with an abnormal interventricular septal movement[6]. This then leads to a regular variation of the diastolic ventricular volume and pressure with each cardiac cycle. The simultaneous performance of the echocardiogram during cardiac catheterization seems to support this hypothesis.

The sign of diastolic pressure alternans during right or left ventricular pressure measurement should alert the operator to the presence of a congenital absence of the pericardium and lead him to the right path of confirming the diagnosis either on CT scan or MRI.

To the author's knowledge, this is the first description of this finding in the literature. Further reports will help in determining whether this observation is a useful sign.

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