CASE REPORT

Ebstein's Disease With Pulmonic Stenosis in an Elderly Man With Three-Vessel Coronary Artery Disease

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ABSTRACT

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KEY WORDS: *Ebstein anomaly; pulmonic stenosis; coronary artery disease; congenital heart disease*

A case is herein presented of a very rare combined congenital anomaly, that of Ebstein's disease and pulmonary valve stenosis, which remained subclinical for a long time until the patient at the age of 65 years developed worsening and crescendo angina due to severe three-vessel coronary artery disease. The patient was successfully managed with surgical repair of the valvular anomalies and with coronary artery bypass grafting of his coronary disease.

INTRODUCTION

Ebstein's disease is an abnormality of the tricuspid valve which is displaced downward toward the right ventricular apex.[1,2] Thus, part of the right ventricle is "atrialized" and located on the atrial side of the tricuspid valve, while the remaining functional right ventricle is small. The tricuspid valve is usually regurgitant. Depending on the severity of the hemodynamic abnormalities in patients with Ebstein's anomaly, determined by the degree of displacement and the functional status of the tricuspid valve, onset of clinical symptoms varies from severe heart failure in the neonate to the absence of symptoms in an adult, in whom it may be an incidental finding. Patients usually have a reduced life expectancy and seldom do they reach an age of >50-60 years without corrective intervention. There are usually other associated congenital anomalies but concomitant occurrence of pulmonary valve stenosis is extremely rare. We present herein a patient with both these disorders which remained subclinical for a long time until the patient at the age of 65 years developed crescendo angina due to severe three-vessel coronary artery disease.

ABBREVIATIONS
CABG = coronary artery bypass grafting
CAD = coronary artery disease
ECG = electrocardiogram
TEE = transesophageal echocardiography
TTE = transthoracic echocardiography

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CASE REPORT

A 65-year-old gentleman was referred for cardiac catheterization by his local physician due to recent re-emergence of anginal symptoms. He was an ex-smoker, hypertensive patient over the past 6 years under therapy and had a history of hyperlipidemia

inadequately controlled with statin therapy. He had a family history strongly positive for coronary artery disease (CAD) with a brother dying suddenly at the age of 42 and another brother having had coronary artery bypass grafting (CABG) at the age of 45 years. He was exempted from his military duty due to a "cardiac murmur".

Three years earlier the patient had developed symptoms of crescendo angina for which he had been submitted to coronary angiography, which had revealed three-vessel CAD with good left ventricular function. He had also undergone right heart catheterization and transesophageal echocardiography (TEE). The initial working diagnosis had been primary pulmonary hypertension versus thrombo-embolic pulmonary disease. Ventilation-perfusion lung scan was of low-probability for pulmonary embolism. Thus, conservative management was recommended at that time for both his 'pulmonary" and cardiac disease.

The patient was placed on anti-anginal medications and remained oligosymptomatic until 3 months prior to this admission, when he developed worsening symptoms of angina. He was hospitalized elsewhere and his medical therapy was intensified with resultant improvement of his symptoms, until he was referred for catheterization.

At **physical examination**, he was an elderly gentleman with mild cyanosis, overweight (height 167 cm, weight 81 kg), in otherwise good general condition. His neck exam was remarkable for a venous pressure of 5-6 cm H_2O with a visible V wave at his jugular venous pulse. His lung examination was normal. There was a palbable left parasternal impulse (right ventricular lift). During cardiac auscultation, S_1 was faint and there was an audible holosystolic murmur grade III/VI at the left parasternal area (tricuspid area). There was a widely split S_2 with a decreased P_2 component. There was also an ejection systolic murmur heard at the right upper sternal border (pulmonary valve are), grade III-IV/VI radiating to the neck. Abdominal examination was remarkable for mild liver enlargement (2 cm below the right costal margin), which was soft and non-tender. There were no lymph nodes palpable. Peripheral pulses were normal, equally and symmetrically palpable. Basic neurological examination was normal with no focal signs.

The **electrocardiogram** showed sinus rhythm at 69 beats per minute, right bundle branch block (RBBB) and right ventricular strain pattern. The PQ interval was at 180 ms and there were q waves in the inferior leads II, III and aVF (Figure 1).

Chest X-ray showed an increased cardiothoracic ratio (56%). **Arterial blood gas** analysis revealed a PH of 7.41, PO_2 62 mmHg, PCO_2 38 mmHg, $HCO_3 = 28$ mmol/l, O_2 saturation 91%. **Laboratory testing** was unremarkable except for a total cholesterol level of 234 mg/dl, HDL-cholesterol 45 mg/dl, LDL-cholesterol 143 mg/dl, and triglyceride level of 230 mg/dl. Electrolytes were normal. **Holter** monitoring did not disclose any supraventricular or ventricular tachyarrhythmias except for isolated ventricular extrasystoles.

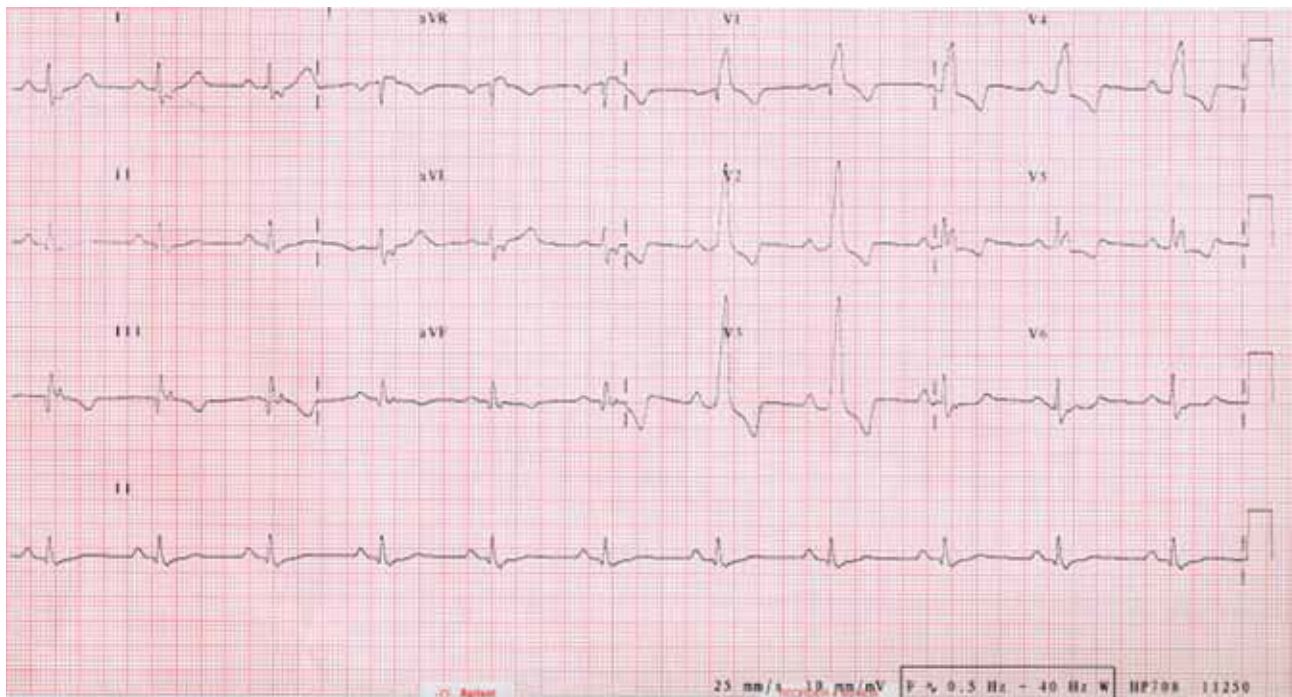The **transthoracic echocardiogram** (TTE) showed normal

FIGURE 1. Twelve-lead electrocardiogram (ECG) with right bundle branch block (RBBB) but no pre-excitation.

left ventricular dimensions with moderate hypokinesia of the infero-basal and infero-apical wall; left ventricular ejection fraction was 50%. The mitral valve was mildly thick and had minimal regurgitation. The aortic valve was tricuspid, mildly thick and calcified but with good opening excursion and with no regurgitation. The main finding was the apical displacement of the septal leaflet of the tricuspid valve and the mast-like appearance of the anterior tricuspid leaflet typical of Ebstein

anomaly with associated severe tricuspid insufficiency grade 3+/4+. The pulmonic valve was thick with significant restriction in its opening and had an hourglass-shaped valve stenosis with hypoplastic main pulmonary artery trunk; maximal systolic pressure gradient was at measured at 74 mmHg (Figures 2-4).

The **transesophageal echocardiogram** (TEE) showed a large waving mast-like anterior leaflet of the tricuspid valve with anomalous attachment at the apex of the right ventricle (Figure 5) and apical displacement of the septal leaflet (Figure 6). The true distal right ventricle was small and hypertrophied (Figure 7) with normal systolic function, while the right atrium was huge, all findings of typical Ebstein's anomaly (Figure 8).[1,2] There was severe tricuspid insufficiency (3+/4+). The pulmonary valve was thick with moderate tubular stenosis
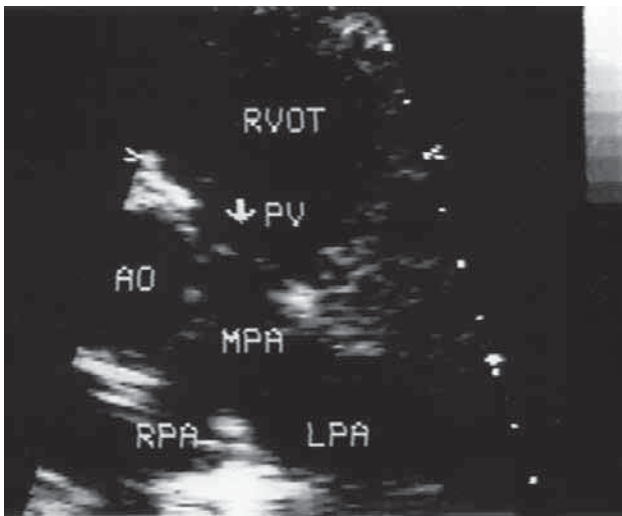

FIGURE 2. Transthoracic echocardiographic short-axis view at the level of the aortic (Ao) valve: pulmonary valve (PV) stenosis with hourglass appearance of a hypoplastic pulmonary artery. LPA = left pulmonary artery; MPA = main pulmonary artery; RPA = right pulmonary artery; RVOT = right ventricular outflow tract.
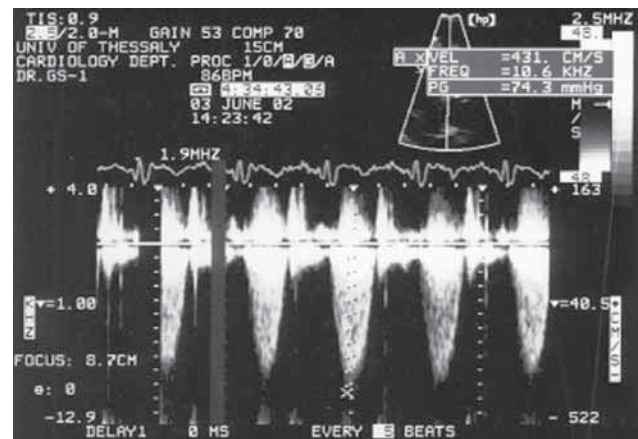

FIGURE 4. Echo-Doppler appearance of the systolic pressure gradient (74 mmHg) across the pulmonary valve.
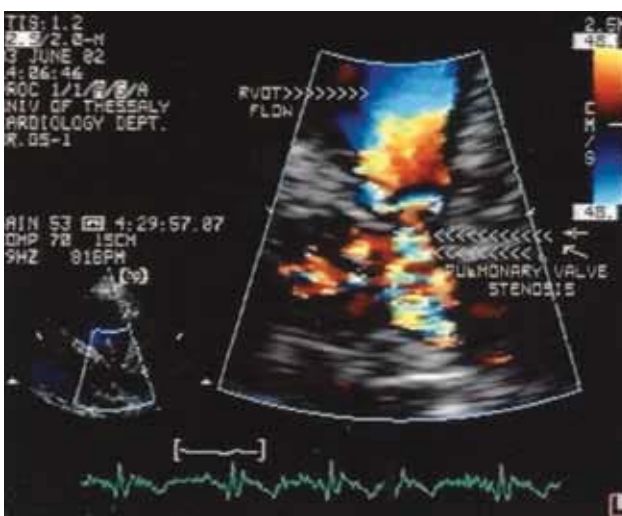

FIGURE 3. Color flow Doppler display of the stenotic pulmonary valve. RVOT = right ventricular outflow tract.
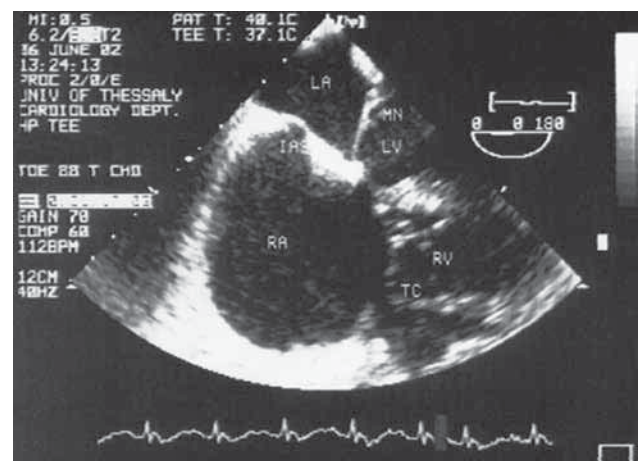

FIGURE 5. Anomalous attachment of the anterior tricuspid leaflet (TC) to the endocardial surface of the apex of the right ventricle (RV). LA = left atrium; LV = left ventricle; RA = right atrium.
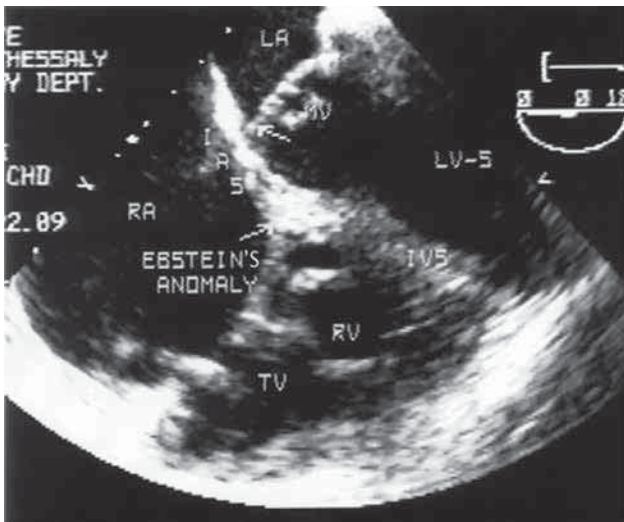
FIGURE 6. Transesophageal echocardiographic 4-chamber view illustrating the apical displacement of the septal tricuspid valve (TV) leaflet, typical of Ebstein's anomaly. IVS = intraventricular septum; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.
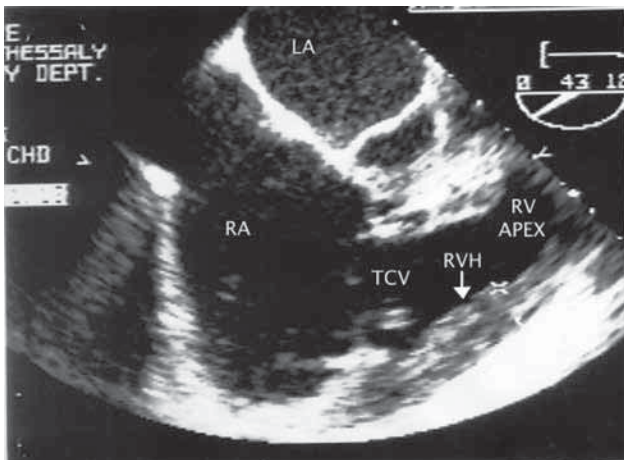

FIGURE 7. Densely hypertrophied right ventricle (RVH) due to co-existence of pulmonic stenosis. LA = left atrium; RA = right atrium; RV = right ventricle; TCV = tricuspid valve.

FIGURE 8. Transesophageal 4-chamber echo view of Ebstein's anomaly. LV = left ventricle; RV = right ventricle.

FIGURE 9. Transesophageal short-axis echo view at the level of the aortic valve (AoV) illustrating the stenosed and thickened pulmonary valve (PVS) without secondary obstruction of the right ventricular outflow tract (RVOT). LA = left atrium; MPA = main pulmonary artery; RA = right atrium.

and the pulmonary arterial trunk was hypoplastic (Figure 9). The right ventricular outflow tract area did not present secondary stenosis. The interatrial septum was intact (Figure 10). The TEE findings were consistent with dual congenital heart disease, i.e. Ebstein anomaly and moderate pulmonic valve stenosis.

During **right heart catheterization**, the systolic pulmonary artery pressure was at 32 mmHg; there was no intra-pulmonary artery gradient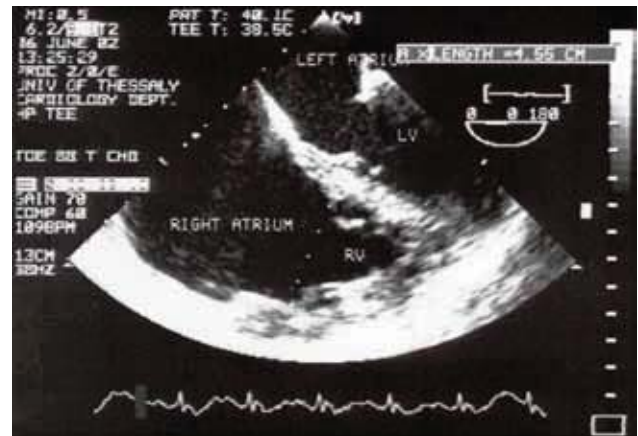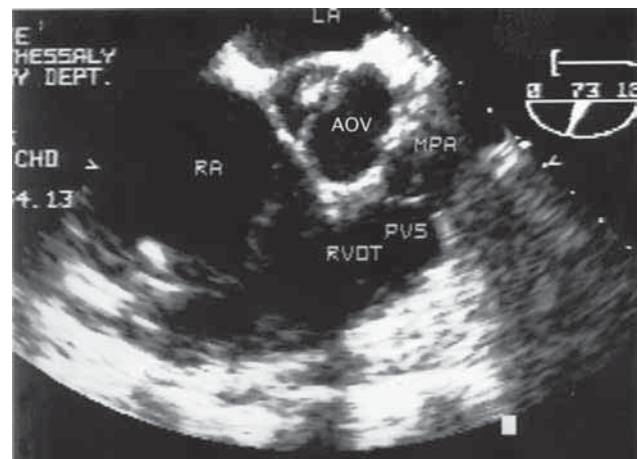 between the trunk and the two branches. The diastolic pulmonary artery pressure was 24 mmHg and the mean pressure was 27 mmHg; the mean wedge pressure was 22 mmHg. The systolic pressure at the outflow tract was 95 mmHg. The systolic pressure of the atrialized part of the right ventricle was 55 mmHg. The A wave of the proximal right atrium was measured at 18 mmHg and the V wave at 25 mmHg. The pressure gradient between the main pulmonary artery and the outflow tract was 65 mmHg (Figures 11A, 11B, 11C).

Coronary angiography revealed a short and patent left main coronary artery and two tandem subtotal occlusions of the left anterior descending coronary artery in the mid segment. The second obtuse marginal branch of the circumflex
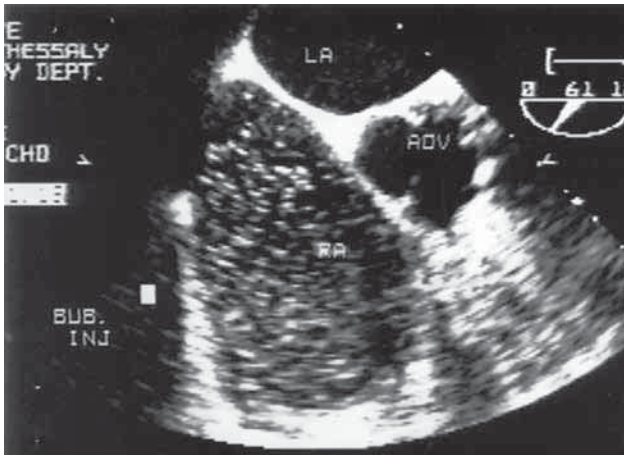
FIGURE 10. Bubble study with infusion of agitated saline, indicating an intact intratrial septum as no bubbles can cross the septum from the right atrium (RA) into the left atrium (LA). AoV = aortic valve.

artery was totally occluded, visualized distally via collaterals. The right coronary artery was a dominant vessel but had total proximal occlusion with mild bridging collaterals and an adequate distal perfusion via collateral circulation from the left coronary artery. Left ventriculography showed moderate hypokinesia of the infero-basal wall but adequate global systolic function of the left ventricle with an ejection fraction of ~55%. The left ventricular pressure was 160/92 mmHg and the aortic pressure was 160/92 mmHg with no systolic gradient across the aortic valve.

In *conclusion*, this was a patient with three-vessel CAD, Ebstein's disease with associated severe tricuspid regurgitation, and a moderate pulmonic valve stenosis (systolic gradient 65 mmHg). The recommendation was for surgical management, to which the patient consented.

During **surgery**, there was visualized an enlarged right atrium, a hypoplastic pulmonary artery and a leftward anatomical axis of the heart. A longitudinal incision of the pulmonary artery was carried out through the level of the pulmonic annulus, followed by commissurotomy of all three commissures and closure was achieved with use of an homologous pericardial patch which was fixed in glurylaldehyde. Then, a right atriotomy was performed, and plication of the base of the septal and posterior tricuspid leaflets was carried out at the level of the true tricuspid annulus with use of pi-shaped

sutures with prolene 4-0 felt pledgets using the De Vega annuloplasty technique.[3] Valve function was then tested and found to be satisfactory. There followed the atriotomy closure with a double layer technique. Subsequently, standard CABG was performed with use of the left thoracic artery to the left anterior descending coronary artery; the left radial artery was anastomosed to the obtuse marginal branch of the left circumflex coronary artery, while the right coronary artery could not be bypassed due to its diffuse disease and absence of a distal segment to target. The patient's post-operative course was uneventful and the patient was discharged home in stable clinical condition.

DISCUSSION

Ebstein's disease is a rare congenital anomaly which occurs in ´1 per 200 000 live births and accounts for <1% of all cases of congenital heart disease.[1,2,4] It involves a malformation of the tricuspid valve and of the right ventricle with downward (apical) displacement of the functional tricuspid annulus. An interatrial communication is present in 80% - 94% of patients with Ebstein's disease.[1] Adults often present with arrhythmias mostly due to accessory pathways, but may also manifest cyanosis, decreasing exercise tolerance, fatigue, or right-sided heart failure. In the presence of an interatrial communication, the risk of paradoxical embolization, brain abscess, and sudden death increases. Additional associated anomalies do occur (e.g., bicuspid aortic valve, pulmonary atresia, subaortic stenosis, coarctation, ventricular septal defect, etc.), but pulmonary valve stenosis is extremely rare. In patients with Ebstein's disease, late onset of clinical symptoms despite severe tricuspid valve displacement and dysfunction is rare.[4,5] Patients usually have a reduced life expectancy and seldom do they reach the age of 50. Only scarcely do they live >70 years without corrective intervention.

The present case illustrates an example of very rare and interesting dual congenital anomaly, that of Ebstein's disease and pulmonic stenosis, which remained subclinical for a long time until the patient developed worsening and crescendo angina due to severe three-vessel CAD. To the best of our knowledge and from reviewing the English literature, we encountered no similar patient with such a triple disease combination, which makes this case unique. The patient was finally managed successfully via surgery,[3,6-8] where both his congenital and acquired diseases were corrected.
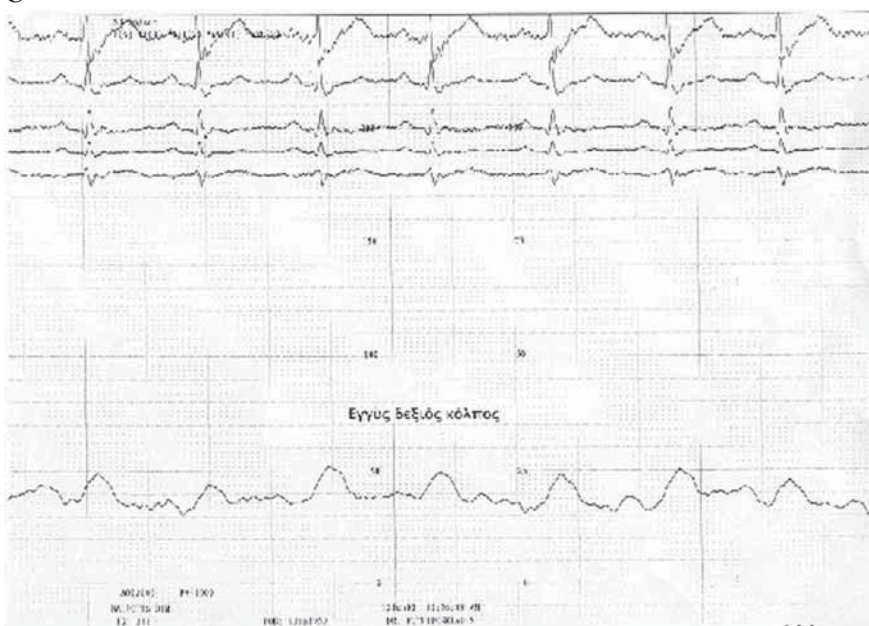
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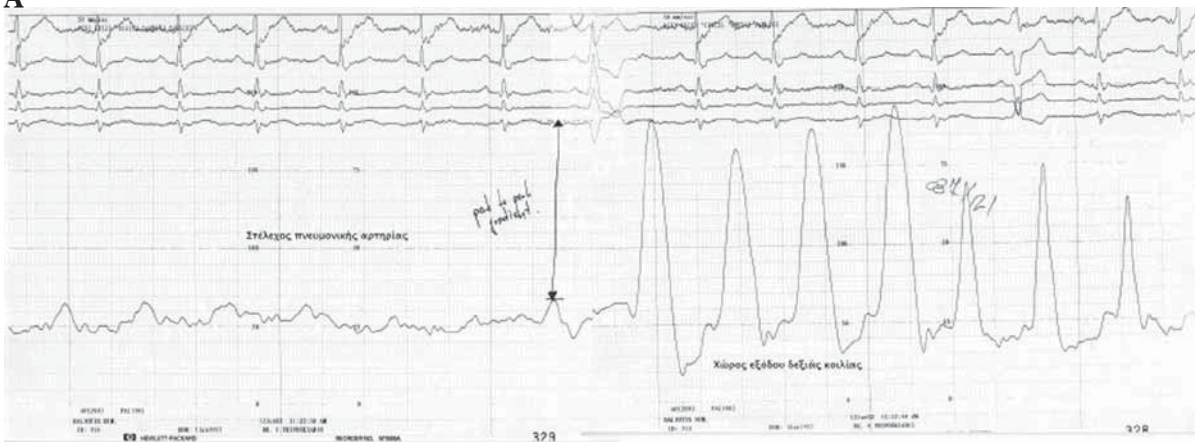
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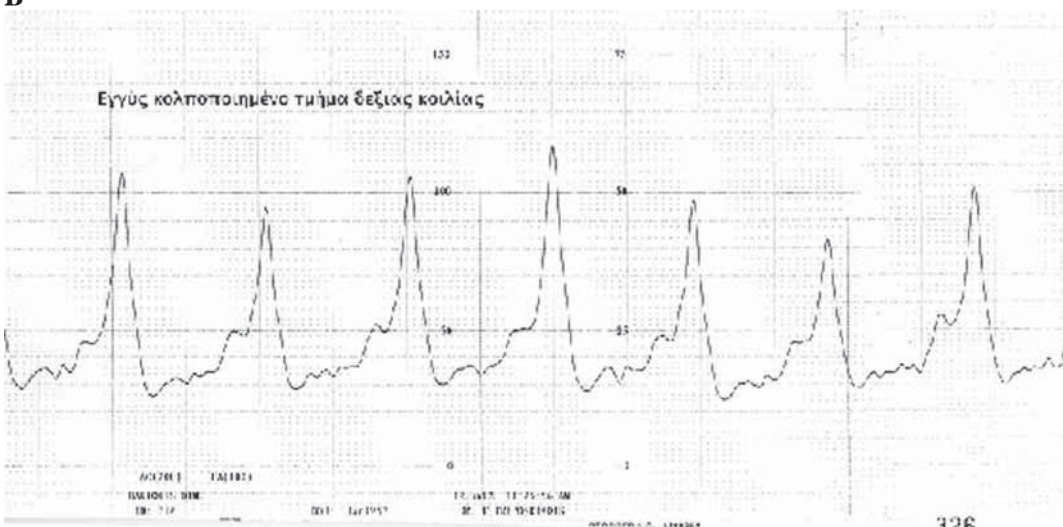
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FIGURE 11. During catheter withdrawal from the main pulmonary artery into the right ventricular outflow tract there is a 65 mmHg pressure gradient recorded (**A**). An intracardiac pressure recording of 55 mmHg is obtained from the atrialized part of the right ventricle (**B**). When the catheter is pulled into the true right atrium the pressure drops to 18 mmHg for the A wave or 25 mmHg for the V wave (**C**).

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