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Aortico-left ventricular tunnel: a rare cause of 'to-and-fro' murmur

Tunel aortalno-lewokomorowy — rzadka przyczyna szmeru skurczowo-rozkurczowego

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Abstract

Aortico-left ventricular tunnel (ALVT) is a very rare congenital paravalvular communication between the aorta and the left ventricle, with fewer than a thousand cases reported to date. Most commonly, the tunnel arises from the right aortic sinus. We herein report the case of a 28 year-old male where a common pouch, which was communicating to the left as well as the right aortic sinus, was draining to the left ventricle. He presented with progressive heart failure where a long diastolic murmur was audible. The patient succumbed to death because of progressive cardiac decompensation before surgical correction could be performed.

This is the first ever report of AVLT with the proximal chamber as a common sac, communicating with both the left as well as the right aortic sinus, was communicating distally to the left ventricle.

Key words: aortico-left ventricular tunnel, congenital heart defect, aortic regurgitation, diastolic murmur, cardiac decompensation

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Introduction

Since first reported as a congenital lesion in 1963, aortico-ventricular tunnel (AVLT) has been a very rare congenital channel between the ascending aorta above the sino-tubular junction proximally to the cavity of the left (90%), or less commonly the right (10%), ventricle distally [1]. This is an exceedingly rare anomaly, with incidence as low as 0.001% of all congenital cardiac diseases [2]. The onset, severity and progression of the disease vary from in-utero foetal demise to asymptomatic adulthood [2]. The onset of heart failure involves interplay between the cross-sectional area of tunnel and the amount of aortic regurgitation. It differs from a ruptured sinus of Valsalva aneurysm in having its proximal origin in the tubular aorta, rather than the aortic sinus, and in passing outside the heart to drain into either ventricle [3]. It is usually extra-cardiac, but rarely may burrow through the intracardiac myocardium to reach the ventricular cavity, a differentiating feature from a coronary-cameral fistula [3]. Associated defects, usually involving the aortic and pulmonary valves, or the proximal coronary arteries in the form of stenosis, regurgitation, dysplasia, atresia, and perforation, have been reported in 50% of cases as the result of hydrodynamic trauma [4].

Case report

A 28 year-old male presented with dyspnoea on exertion [New York Heart Association (NYHA) functional class IV] for the past six months. The patient had been diagnosed as

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a case of aortic valve disease with aortic regurgitation three years previously. On clinical examination, blood pressure in the upper and lower limbs was 110/40 and 160/40 mm Hg respectively. Pulse rate was 98/min, regular, bounding, with no radio-radial or radio-femoral delay, and all pulses were palpable. His jugular venous pressure was raised 5 cm above the sternal angle. Cardiac apex was located in the 6 left parasternal region, 3 cm lateral to mid clavicular line which was hyperdynamic in character. On auscultation, S1 was soft, S2 was loud with a loud P2 component, and S3 was heard. A loud 'to-and-fro' murmur, with systolic and diastolic thrills radiating over the entire precordium, and bounding pulses indicating rapid aortic run-off was noted. A long early diastolic murmur along the left 3rd parasternal area and an ejection systolic murmur of grade IV/VI were also heard. Electrocardiogram showed evidence of left ventricular hypertrophy with ST-T changes in lateral precordial leads. Chest X-ray revealed cardiomegaly. Two-dimensional transthoracic echocardiography in apical four-chamber view revealed an aortic regurgitation jet (Figure 1A). In parasternal long-axis (PLAX) and short axis views (SAX), a common sac was seen communicating with the right as well as the left aortic sinuses, which was connecting with the left ventricle along with a jet of valvular aortic and mitral regurgitation (Figures 1B, 2, 3). Aortic valve was tricuspid. Left ventricular dimensions were enlarged, with systolic dysfunction (ejection fraction 30%). Three--dimensional echocardiography (Phillips i33) showed the

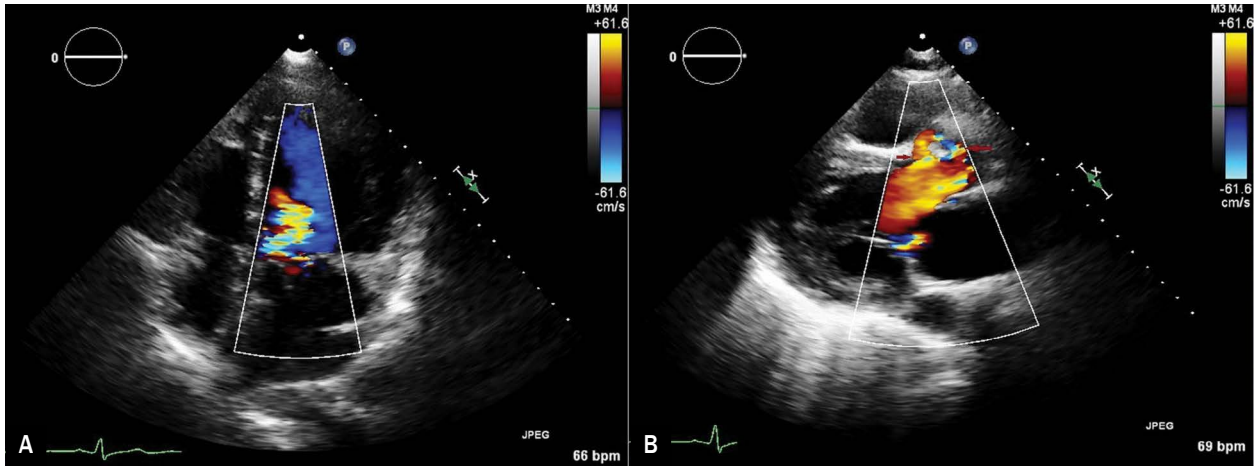

Figure 1A, B. Two-dimensional transthoracic echocardiography in apical four-chamber view revealed aortic regurgitation jet (**A**); parasternal long-axis view showing two jets (red arrow; **B**) communicating with left and right aortic sinus opening into left ventricle along with aortic and mitral regurgitation

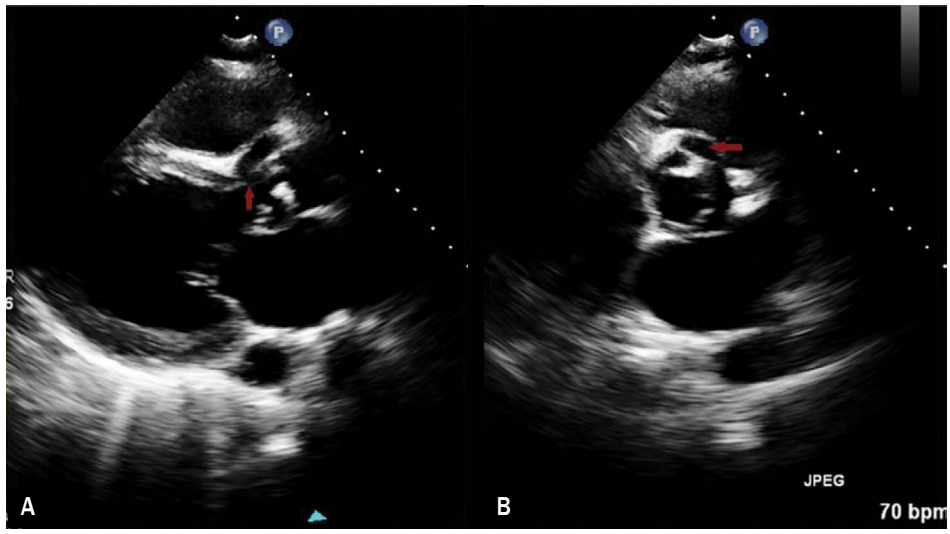

Figure 2A, B. PLAX view showing aortic-left ventricular tunnel (red arrow; **A**); short axis view showing a common sac (red arrow; **B**)

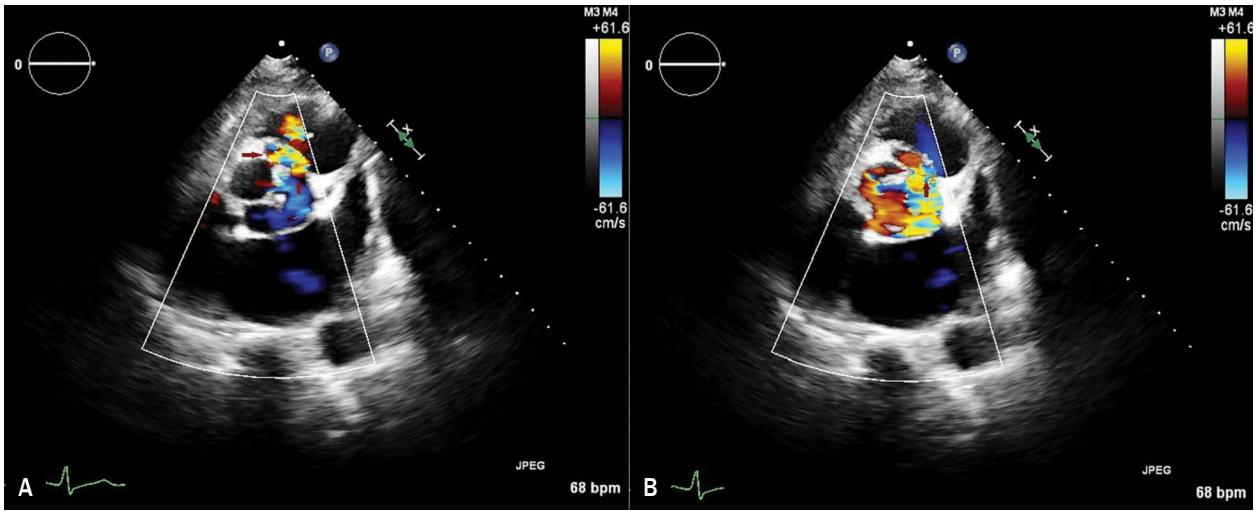

Figure 3A, B. Short axis view showing a common sac communicating with right (red arrow; **A**), and left aortic sinus (red arrow; **B**)

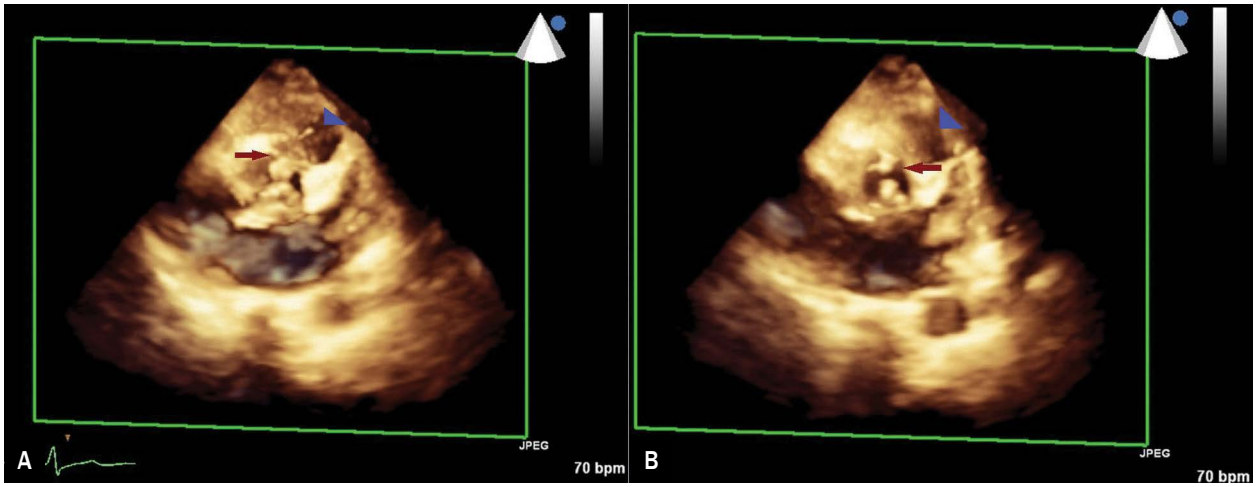

Figure 4A, B. Short axis view showing a proximal common chamber (blue arrowhead) communicating with right (red arrow; **A**), and left aortic sinus (red arrow; **B**)

proximal common chamber communicating with both the right as well as the left aortic sinuses (Figure 4). Multidetector computed tomography (MDCT) revealed AVLT where the detail of the abovementioned findings was amplified in volume rendered reconstruction (Figures 5, 6). Cardiac catheterisation with an angiographer was deferred because of haemodynamic instability. Before it could take place, the patient died due to progressive failure.

Discussion

The exact aetiology of aortico-ventricular tunnel is unknown. Probably it results from a combination of maldevelopment and abnormal separation of the cushions which give rise to the pulmonary and aortic roots [3]. Such patients have severe aortic regurgitation (AR) and present with heart failure. Mostly patients became symptomatic in infancy. Symptoms can present at any time but rarely beyond the second decade. AVLT must be distinguished from the lesions producing rapid run-off of blood from the aorta causing cardiac failure such as a sinus of Valsalva fistula, truncus arteriosus with regurgitation, aorto-pulmonary window, ventricular septal defect with aortic regurgitation, patent ductus arteriosus, coronary-cameral fistula, aortic stenosis with regurgitation, and tetralogy of Fallot with absent pulmonary valve, to name but a few. These can be reliably differentiated with the help of echocardiography, MDCT, cardiac catheterisation and magnetic resonance imaging. Echo is the diagnostic investigation of choice because it can demonstrate aortic and ventricular openings and the

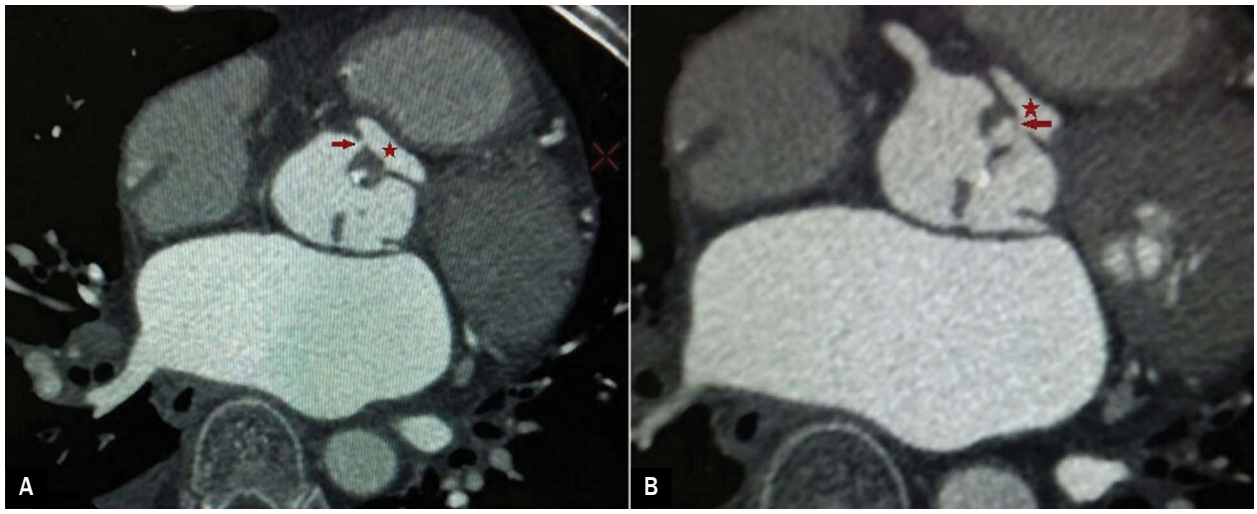

Figure 5A, B. Multidetector computed tomography (MDCT) in axial plane revealed a common proximal chamber (red asterix) communicating with right (red arrow; **A**), and left aortic sinus (red arrow; **B**)

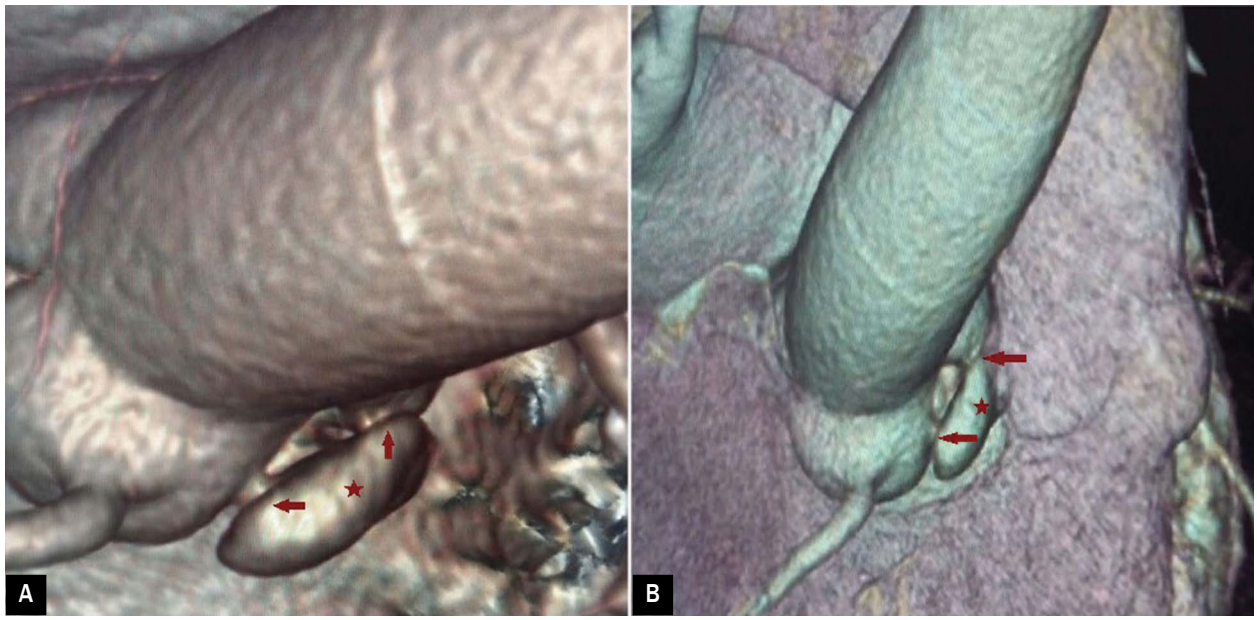

Figure 6. Volume rendered reconstruction (VR) showing a common proximal chamber (red asterix) communicating with right (red arrow; **A**), and left aortic sinus (red arrow; **B**)

tunnel. Colour Doppler can show the flow pattern. Cardiac catheterisation is required for associated lesions and to understand the coronary artery anatomy.

The ostium of a coronary artery may lie within an aortico-ventricular tunnel, though it arises from the tubular part of the aorta. Variable involvement of the ostia in the form of atresia of the left or right have both been reported [5, 6]. Surgical interruption of the tunnel should be undertaken without delay, even in asymptomatic patients, to prevent left ventricular dysfunction, although spontaneous closure has been reported in one patient. There have been some observations of an occasional asymptomatic patient with a very small (< 2 mm) aorto-left ventricular tunnel [7]. Many patients already have associated aortic regurgitation, as this is a major long-term complication after surgery, requiring valve replacement in 50% of the cases [8, 9]. The causes of acquired aortic incompetence are multiple, and can include unsupported aortic cusp, progressive aortic root dilatation, perforations in the leaflet due to hydrodynamic trauma, or a primary valvular defect [4]. In adult survivors, progressive aneurysmal dilatation of the aortic root is an important issue. Hence, early correction is indicated, not

only to prevent heart failure but also to prevent progression of damage to the aortic valve [10]. Furthermore, it has been observed that patients who were operated within the first six months of life had documented normalisation of left ventricular size and function, although the aetiology of progressive dilatation of aortic root and development of valvular aortic regurgitation noted in patients who underwent tunnel closure in early infancy is yet unknown [11].

Surgical intervention consists of closing the tunnel such that the aortic valve is supported, the coronary circulation is not compromised, and the left or right ventricular outflow obstruction is prevented or relieved. In most cases of AVLT, this is achieved by transaortic patch closure of the aortic end, and placement of a second patch through the tunnel itself to close the ventricular orifice and support the aortic valve. Both the ends need to be closed. If the ventricular end of an aorto-left ventricular tunnel is not closed, residual high pressure in the blind-ending pouch may compress the right ventricular outflow [4]. Transcatheter closure of a tunnel to the left ventricle with an Amplatzer duct occluder is another modality that has been reported in two patients [12, 13]. This should not be done in patients with advanced ventricular dysfunction where the risk outweighs the benefit. Our case is unique, extremely rare and here being reported for the first time as the proximal chamber was connecting with both the left and right aortic sinuses. Also, the patient had remained asymptomatic until his third decade of life.

Conflict(s) of interest

The authors declare no conflict of interest.

Streszczenie

Tunel aortalno-lewokomorowy (AVLT) to występujące bardzo rzadko wrodzone okołozastawkowe połączenie między aortą a lewą komorą. Dotychczas opisano mniej niż 1000 przypadków tej wady serca. Ujście tunelu znajduje się najczęściej w prawej zatoce aortalnej. W artykule przedstawiono przypadek 28-letniego mężczyzny, u którego wspólna kieszonka komunikująca się zarówno z prawą, jak i lewą zatoką aortalną uchodziła do lewej komory. Chory zgłosił się z narastającą niewydolnością serca. W badaniu osłuchowym był słyszalny długi szmer rozkurczowy. Pacjent zmarł z powodu postępującej dekompensacji serca, zanim było możliwe wykonanie chirurgicznego zabiegu naprawczego.

Jest to pierwszy w historii opis AVLT z komorą proksymalną jako wspólną kieszonką, komunikującą się zarówno z prawą, jak i lewą zatoką aortalną uchodzącą do lewej komory.

Słowa kluczowe: tunel aortalno-lewokomorowy, wrodzona wada serca, niedomykalność aortalna, szmer skurczowy, dekompensacja krążeniowa

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