Right atrial inflow obstruction of the inferior vena cava due to atrial septal aneurysm and an elongated Eustachian valve

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We describe a patient with oedema of the legs and ascites due to right atrial (RA) inflow obstruction, caused by atrial septal aneurysm (ASA) and an elongated Eustachian valve (EV). Both structural abnormalities created a narrow inflow channel to the free RA cavity. RA inflow obstruction is usually related to constrictive pericardial disease and pericardial tamponade. Other cardiac causes of RA inflow obstruction are rare and include lipomatous hypertrophy of interatrial septum, tumour or thrombus in the right atrium (RA). Reports, describing inflow obstruction in the RA as a consequence of an elongated EV or an ASA, are extremely rare.

KEYWORDS
Right atrial inflow obstruction; Inferior vena cava; Elongated Eustachian valve; Atrial septal aneurysm; Transesophageal echocardiography; Congenital heart disease

Case presentation
A 71-year-old female, with a medical history of transient ischaemic attack, arterial hypertension, and monoclonal IgA gammopathy was referred to our department by her physician.

During 6 months, the patient progressively developed ascites, pitting oedema of the legs and an increase of 12 kg in weight. She was fatigued. There was no orthopnea.

A work-up to exclude an intra-abdominal etiology for the fluid retention was already performed in the department of internal medicine. Tumour markers were negative. There was mild hypoalbuminemia but no albuminuria. Axial computed tomography of the abdomen confirmed hepatomegaly and ascites, but could not reveal intra-abdominal masses. There was no compression of the inferior vena cava (IVC). The analysis of the peritoneal fluid confirmed a transudate. Treatment with furosemide was initiated and the patient was referred to the cardiologist for further examination.

Upon physical examination, the blood pressure was 135/75 mmHg at a pulse rate of 80 beats per minute. Central venous pressure was estimated to be within normal limits. Auscultation of the heart and lungs was unremarkable. There was ascites and pitting oedema of the legs.

Transthoracic echocardiography (TTE) showed normal dimensions of the four chambers. There was normal left ventricular systolic function and impaired diastolic relaxation. Mild regurgitations of atrioventricular valves were documented. Continuous wave-pulsed Doppler measured a maximal tricuspid regurgitation (TR V_max 2.15 m/s) gradient of 18 mmHg. The IVC was dilated without diameter changes during respiration. A floppy interatrial septum (IAS) was seen.

To further evaluate the IAS and the inferior vena cava venous inflow in the right atrium (RA), a transesophageal echocardiography (TEE) was performed. This examination showed an atrial septal aneurysm (ASA), narrowing the right atrial (RA) inflow tract of IVC. The Eustachian valve (EV) was elongated. Both structures formed a thin channel with flow acceleration to the RA cavity documented by colour Doppler (Figures 1 and 2, see Supplementary data online, Videos 1–3).

A balloon dilatation was considered too risky given the anatomical complexity of the lesion, including a channel created by two congenital abnormalities. A stent implantation in the channel seemed to be contraindicated as it was thought to be impossible to obtain a reliable fixation of the stent. Surgical relief of the inflow obstruction was however refused by the patient.
Nevertheless, the patient agreed with a cardiac catheterization. Measurements with the Swan–Ganz catheter documented normal PCW, pulmonary, right ventricular, RA and superior cava venous pressures. There was absence of shunting as O₂ saturation jumps could not be demonstrated. The pullback curve from the RA to the IVC demonstrated a peak-to-peak gradient of 16 mmHg (Figure 3), confirming RA inflow obstruction. Venography of the IVC excluded external compression on the IVC. Angiographic imaging of RA inflow (catheter in the IVC just below the diaphragm) demonstrated passage of contrast through a sickle-shaped tract in the RA before spreading in the RA cavity (Figure 4, see Supplementary data online, Videos 4 and 5).

The dose of furosemide was augmented with substantial clinical improvement. However, mild oedema of the legs was still present. Echocardiographic re-evaluation demonstrated a dilated IVC and hepatic venous system without thrombi (Figure 5).

Discussion

In this report, we describe an adult with two congenital cardiac abnormalities: ASA and an elongated EV. Both structures formed a narrow sickle-shaped tract, causing RA inflow obstruction of the IVC.

In most cases, RA inflow obstruction is caused by constrictive pericardial disease and pericardial tamponade. Other cardiac etiologies of RA inflow obstruction are rare and include lipomatous hypertrophy of the IAS,¹ tumour²,³ or thrombus⁴ in the RA.

Of these, lipomatous hypertrophy of the IAS is the most frequently reported one. EV, causing RA inflow obstruction of the IVC, is extremely rare. To the best of our knowledge, this is the third case report describing RA inflow obstruction due to an EV.⁵,⁶

This case demonstrates the value of TEE in the work-up of not-well-understood RA inflow obstruction of the IVC. TTE excluded left ventricular dysfunction, pulmonary hypertension, pulmonary and tricuspid valve disease, right ventricular dysfunction, pericardial tamponade as etiology of the swollen legs. The Doppler flow patterns did not fit with constrictive pericarditis. The above examinations substantially short tailed the list of causes of the swollen legs. The remaining differential diagnosis included local pericardial constriction causing RA inflow obstruction, external RA compression in the region of the inflow in the IVC, intraluminal obstruction in the upper part of the IVC—the so-called Budd-Chiari syndrome (BCS)—or an intra-cardiac inflow obstruction of the IVC. Finally, RA inflow obstruction of the IVC due to a thin sickle-shaped
channel, formed by ASA and elongated EV, was diagnosed by TEE.

This case shows striking clinical and pathophysiological similarities with the BCS, a well-known clinical entity in hepatology. BCS is an obstruction of the hepatic veins or IVC above the entrance of the hepatic veins or both. This syndrome is most commonly caused by a hepatocellular carcinoma. BCS also occurs as complication after liver transplantation. BCS can also be caused by a membrane in the IVC, which is predominately diagnosed in the Asian population. In this case, the medical problem could be considered as a variant of the BCS, with the obstruction located just above the IVC in the RA, instead of superior part of the IVC. The venography of the IVC excluded BCS (Figure 5).

As mentioned in the case presentation, our patient refused to consider surgical relief of RA inflow obstruction. The symptoms could be reduced by initiating treatment with diuretics. Given the lack of literature on the natural history of inflow obstruction due an elongated EV and ASA, no predictions can be made on the expected symptom-free period.

However, studies on BCS due to a membrane in the IVC, comparing medical treatment with stenting of lesion may provide some information. In the series of Lee et al.7 of 28 patients with BCS due to membranes in IVC or suprahepatic veins, the medical treatment remained effective only in a limited group of 6 (21.4%) of the 28 patients. The results in invasive group were excellent.
These figures suggest an unfavourable outcome without surgical relief. In addition it remains uncertain why first symptoms occurred at the age of 71 years.

In conclusion, we report a patient with right atrial inflow obstruction of the IVC, caused by the combination of an elongated EV and ASA. IVC outflow obstruction due an elongated EV is extremely rare and to the best of our knowledge, this is the third case described in the medical literature.

**Supplementary data**

Supplementary data are available at *European Journal of Echocardiography* online

**References**