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A Rare Congenital Triad: Ebstenoid Tricuspid Valve, Giant Right Atrial Aneurysm, and Atrial Septal Defect

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Case Report

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Abstract

Background

Ebstein anomaly is a rare congenital malformation of the tricuspid valve, accounting for less than 1% of all congenital heart diseases. Ebstenoid anomaly is an uncommon morphological variant characterized by abnormal tricuspid valve leaflet morphology without the classical apical displacement of the septal leaflet seen in Ebstein anomaly. The coexistence of an Ebstenoid anomaly, giant right atrial aneurysm (RAA), and atrial septal defect (ASD) is exceptionally rare.

Case Presentation:

A 15-year-old girl presented with progressive exertional dyspnoea (New York Heart Association [NYHA] class II) and intermittent palpitations. Chest radiography demonstrated marked cardiomegaly. Transthoracic echocardiography revealed a giant right atrium, severe low-pressure tricuspid regurgitation, an ostium secundum ASD, and an Ebstenoid tricuspid valve with an elongated anterior leaflet and tethered septal leaflet, without apical displacement or right ventricular atrialization. Four-dimensional echocardiography and cardiac computed tomography confirmed the anatomical findings. Cardiac catheterization demonstrated pulmonary artery pressures approximately one-third of systemic pressure, confirming operability. The patient underwent successful tricuspid valve repair, ASD closure, and right atrial reduction atrioplasty. At one-year follow-up, she remained asymptomatic (NYHA class I) with preserved right ventricular function and only trivial residual tricuspid regurgitation.

Conclusion

This case describes an exceptionally rare combination of Ebstenoid tricuspid valve anomaly, giant right atrial aneurysm, and an ostium secundum ASD. Multimodality imaging was essential for accurate anatomical delineation and surgical planning. Early surgical correction resulted in excellent clinical and echocardiographic outcomes, underscoring the importance of timely intervention in complex congenital tricuspid valve anomalies.

Introduction

Ebstein anomaly is an uncommon congenital malformation caused by incomplete delamination of the septal and posterior tricuspid valve leaflets, leading to tricuspid regurgitation, atrialization of the right ventricle, and progressive right-sided chamber enlargement [1–3]. Associated atrial septal defect (ASD) or patent foramen ovale occurs in approximately 50–80% of patients and contributes to chronic right-sided volume overload and arrhythmias [2, 4].

The term Ebstenoid anomaly has been used to describe congenital tricuspid valve abnormalities resembling Ebstein anomaly but lacking the classical apical displacement of the septal leaflet. Because this entity is rarely reported, its natural history and optimal surgical management remain poorly defined [5, 6].

Massive right atrial enlargement secondary to severe tricuspid regurgitation is common; however, development of a true giant right atrial aneurysm is exceedingly uncommon [5, 6]. Such patients are at increased risk of atrial arrhythmias, thromboembolic events, progressive right ventricular dysfunction, and heart failure. Modern multimodality imaging including two-dimensional, three-dimensional and four-dimensional echocardiography, cardiac computed tomography (CT), and cardiac magnetic resonance imaging (MRI) plays a pivotal role in defining anatomy and planning surgery [7, 8].

We describe a rare case of a giant right atrial aneurysm secondary to an Ebstenoid tricuspid valve anomaly associated with a secundum ASD in an adolescent successfully treated with tricuspid valve repair, ASD closure, and right atrial reduction atrioplasty.

Case Presentation

A 15-year-old girl presented with a five-year history of progressively worsening exertional dyspnoea and intermittent palpitations. Her functional status had gradually deteriorated from New York Heart Association (NYHA) class I to class II. There was no history of cyanosis, syncope, rheumatic fever, recurrent respiratory tract infections, or previous cardiac intervention.

On physical examination, a hyperdynamic right ventricular impulse was noted. Cardiac auscultation revealed a grade III/VI pansystolic murmur at the left lower sternal border that increased with inspiration, consistent with severe tricuspid regurgitation. There was no peripheral oedema, cyanosis, jugular venous distension, or hepatomegaly.

Electrocardiography demonstrated sinus rhythm with right atrial enlargement, right-axis deviation, and incomplete right bundle branch block. Chest radiography showed marked cardiomegaly with prominence of the right cardiac border, consistent with severe right atrial enlargement, and a cardiothoracic ratio of 75% (Fig. 1).

Transthoracic echocardiography revealed a massively dilated right atrium measuring **9.0 × 9.8 cm** and a dilated right ventricle measuring **4.9 × 5.0 × 6.5 cm**, with preserved right ventricular systolic function (Figs. 2 and 3). The tricuspid valve demonstrated an elongated anterior leaflet and a tethered septal leaflet without significant apical displacement, consistent with an Ebstenoid anomaly. Severe low-pressure tricuspid regurgitation and an ostium secundum atrial septal defect with left-to-right shunting were identified (Video 01 and 02). The estimated right ventricular systolic pressure was **46 mmHg** (excluding right atrial pressure), suggesting moderate pulmonary hypertension (Fig. 4).

Four-dimensional echocardiography provided detailed assessment of tricuspid valve morphology and leaflet coaptation (Fig. 05). Cardiac computed tomography confirmed a giant aneurysmal right atrium measuring **9.3 × 9.4 cm** (area **78 cm²**) without intracavitary thrombus and excluded associated congenital cardiovascular anomalies (Fig. 5). Cardiac catheterization demonstrated pulmonary artery pressure approximately one-third of systemic pressure, indicating favourable pulmonary vascular resistance and confirming operability.

Following multidisciplinary heart team discussion, the patient was scheduled for surgical correction because of progressive symptoms, severe tricuspid regurgitation, giant right atrial aneurysm, and the anticipated long-term risks of atrial arrhythmias, thromboembolism, and right ventricular dysfunction.

Surgical Management

The patient underwent elective surgery through median sternotomy under cardiopulmonary bypass. Intraoperative inspection revealed a massively aneurysmal right atrium with a markedly thinned wall. The tricuspid valve demonstrated an elongated anterior leaflet and tethered septal leaflet producing severe malcoaptation, while atrialization of the right ventricle was minimal.

Native tricuspid valve repair was performed by mobilization of the anterior leaflet, release of tethering where feasible, and annuloplasty to restore leaflet coaptation. The secundum ASD was closed surgically, and excess aneurysmal right atrial tissue was excised followed by reduction atrioplasty. Intraoperative transoesophageal echocardiography confirmed satisfactory valve competence, intact ASD closure, marked reduction in right atrial size, and preserved right ventricular function.

The postoperative course was uneventful. The patient remained in sinus rhythm without conduction disturbances. Serial echocardiography demonstrated trivial residual tricuspid regurgitation, preserved right ventricular function, and no residual atrial shunt. She was discharged in stable condition.

At one-year follow-up, the patient remained asymptomatic (NYHA class I). Echocardiography showed sustained reduction in right atrial size, stable tricuspid valve repair, preserved right ventricular function, and no recurrence of aneurysmal dilatation.

Discussion

Ebstein anomaly encompasses a broad spectrum of congenital tricuspid valve abnormalities resulting from abnormal embryological delamination of the valve leaflets [1, 2]. The Ebstenoid variant is considerably less common and is characterized by dysplastic leaflet morphology without the typical apical displacement required for the diagnosis of classical Ebstein anomaly [5]. Recognition of this distinction is important because surgical repair should be individualized according to valve morphology.

Our patient presented with an exceptionally rare combination of an Ebstenoid tricuspid valve anomaly, giant right atrial aneurysm, and secundum ASD. Although right atrial enlargement frequently

accompanies severe tricuspid regurgitation, aneurysmal enlargement of this magnitude has only rarely been described. To our knowledge, only a few cases of anterior displacement of the tricuspid valve anterior leaflet or displacement of all three tricuspid valve leaflets have been described in the literature (5,6). Chronic right-sided volume overload due to severe tricuspid regurgitation, compounded by left-to-right shunting across the ASD, likely contributed to progressive aneurysmal dilatation.

Multimodality imaging was essential for diagnosis. Transthoracic echocardiography established the presence of severe tricuspid regurgitation and abnormal valve morphology, whereas four-dimensional echocardiography provided superior assessment of leaflet motion and coaptation. Cardiac CT accurately delineated the extent of aneurysmal enlargement and excluded associated vascular abnormalities, facilitating surgical planning [7, 8].

The principal goals of surgery were restoration of tricuspid valve competence, elimination of intracardiac shunting, and reduction of the massively dilated right atrium. Contemporary management of Ebstein anomaly increasingly favours valve repair over replacement because preservation of the native valve provides superior long-term right ventricular function and avoids prosthesis-related complications, particularly in young patients [9, 10]. Right atrial reduction atrioplasty is an effective adjunctive procedure that decreases atrial volume, reduces wall stress, improves cardiac geometry, and may reduce the future risk of atrial arrhythmias [6].

Early surgical intervention before irreversible right ventricular dysfunction develops is associated with excellent functional recovery and improved long-term outcomes [9, 10]. Our patient demonstrated marked symptomatic improvement, stable valve function, and preserved right ventricular performance one year after surgery.

This case emphasizes the importance of multidisciplinary management involving pediatric cardiologists, cardiac surgeons, imaging specialists, and anaesthesiologists. Individualized surgical planning based on comprehensive anatomical assessment resulted in an excellent early outcome despite the complexity of the congenital abnormalities.

The principal limitation of this report is the relatively short duration of follow-up. Continued surveillance is required to evaluate long-term durability of valve repair, right ventricular function, and the occurrence of late arrhythmias.

Conclusion

Giant right atrial aneurysm secondary to an Ebstenoid tricuspid valve anomaly associated with a secundum ASD is an exceptionally rare congenital cardiac lesion. Comprehensive multimodality imaging accurately defined the complex anatomy and guided successful surgical repair. Combined tricuspid valve repair, ASD closure, and right atrial reduction atrioplasty resulted in excellent clinical and echocardiographic outcomes. Early recognition and timely intervention are essential to prevent progressive right-sided heart failure and arrhythmic complications.

Learning objectives:

1. **Morphological Differentiation**: Differentiate an Ebstenoid tricuspid valve anomaly (abnormal leaflet morphology without classical apical displacement or ventricular atrialization) from typical Ebstein anomaly.
2. **Diagnostic Evaluation**: Explain the role of multimodality imaging (4D echocardiography, cardiac CT) and cardiac catheterization in assessing anatomical details, pulmonary hemodynamics, and surgical operability.
3. **Surgical Management & Outcomes**: Identify the key surgical procedures (tricuspid valve repair, ASD closure, and right atrial reduction atrioplasty) required to achieve long-term functional recovery and symptom relief.

Abbreviations

RAA
Right atrial aneurysm
ASD
Atrial septal defect
NYHA
New York Heart Association
ECG
Electrocardiography
2D ECHO
Two dimensional Echocardiography
CT
Computed tomography
MRI
Magnetic resonance imaging (MRI)

Declarations

Consent to publish:

Written and informed consent was obtained from the parents of the patient

Author Contribution

UDK: Conceptualization; Methodology; Supervision; Writing—original draft; Writing—review and editing.. SA: Validation; Writing—original draft; Writing—review and editing. ND: Writing—review and editing. All authors reviewed the paper and approved the final version of the manuscript.

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Figures

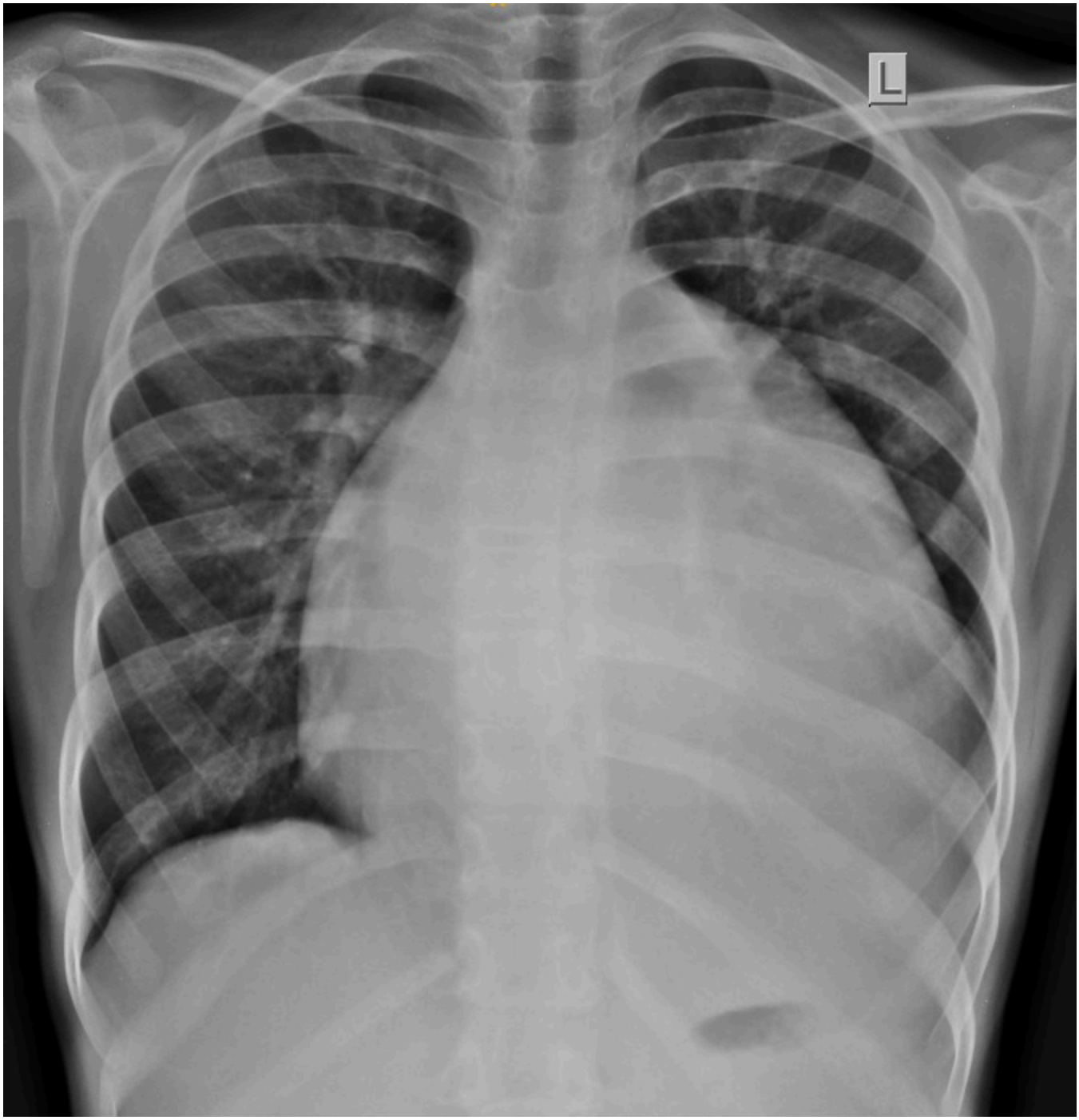

Figure 1

Chest radiograph (posteroanterior view)demonstrating marked cardiomegaly with prominent right atrial enlargement, elevation of the right ventricular apex, and a cardiothoracic ratio of **75%**.

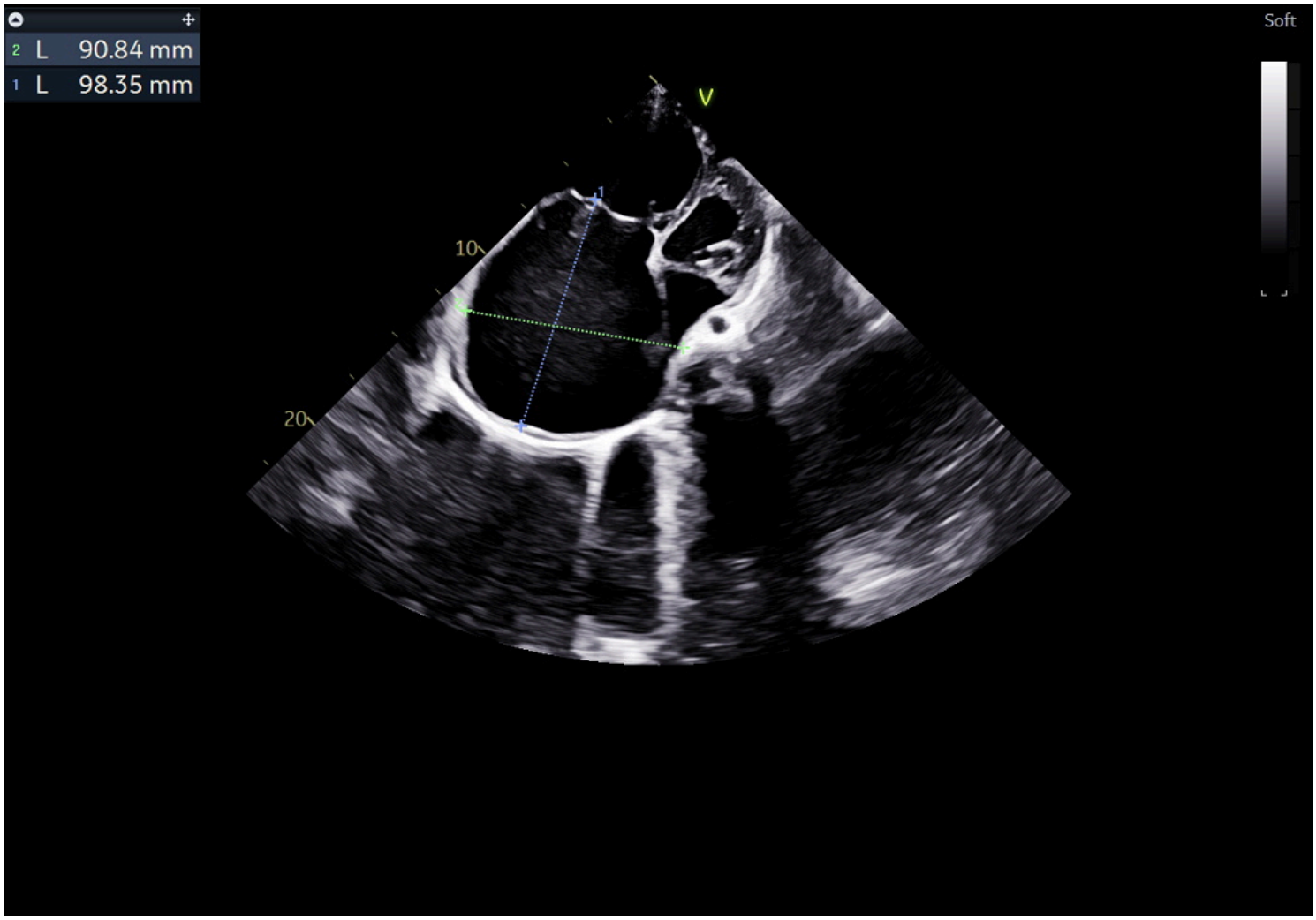

Figure 2

Transthoracic echocardiography (apical four-chamber view) showing a massively dilated right atrium measuring **9.0 × 9.8 cm**.

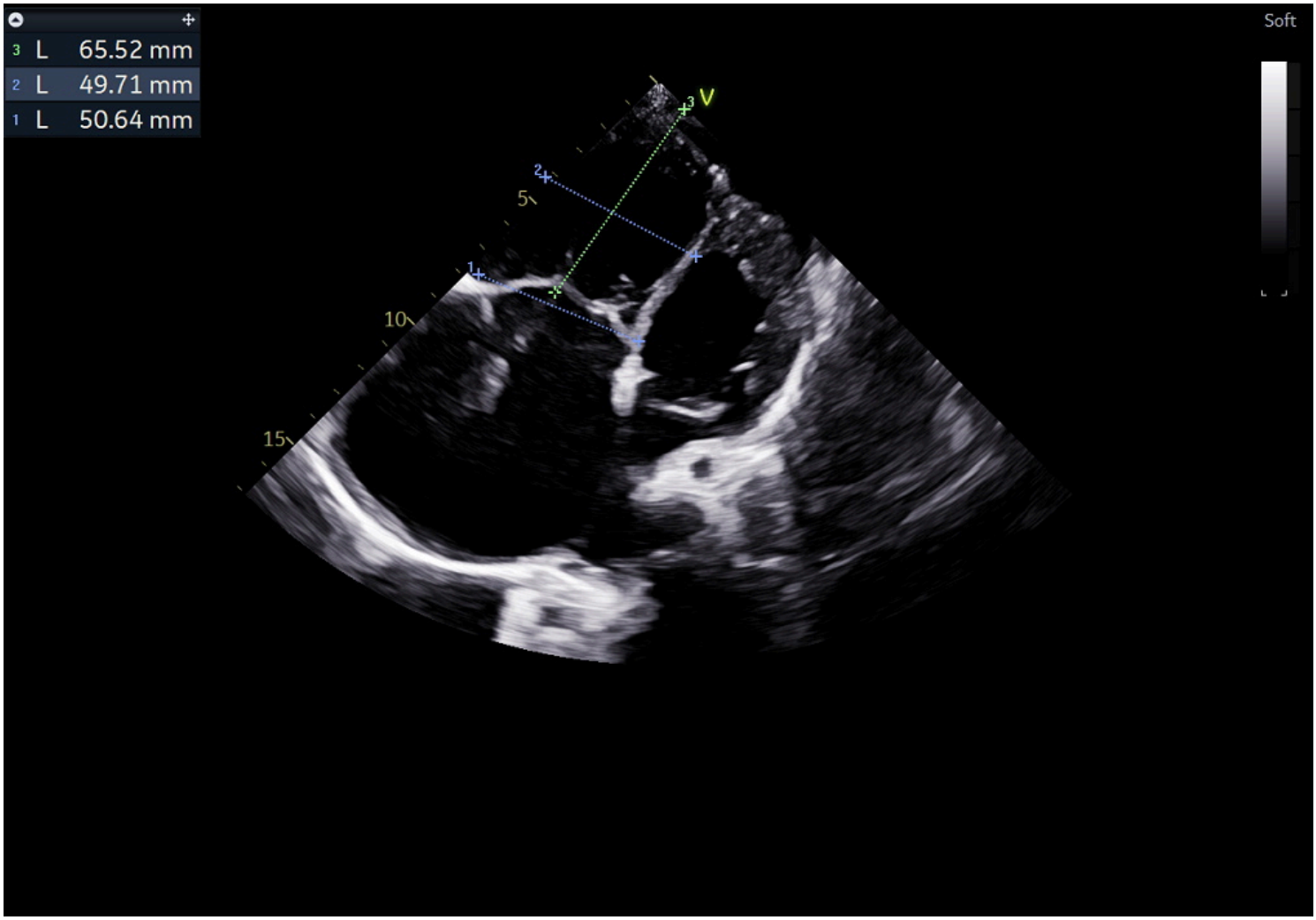

Figure 3

Transthoracic echocardiography (apical four-chamber view) demonstrating a dilated right ventricle measuring **4.9 × 5.0 × 9.8 cm**.

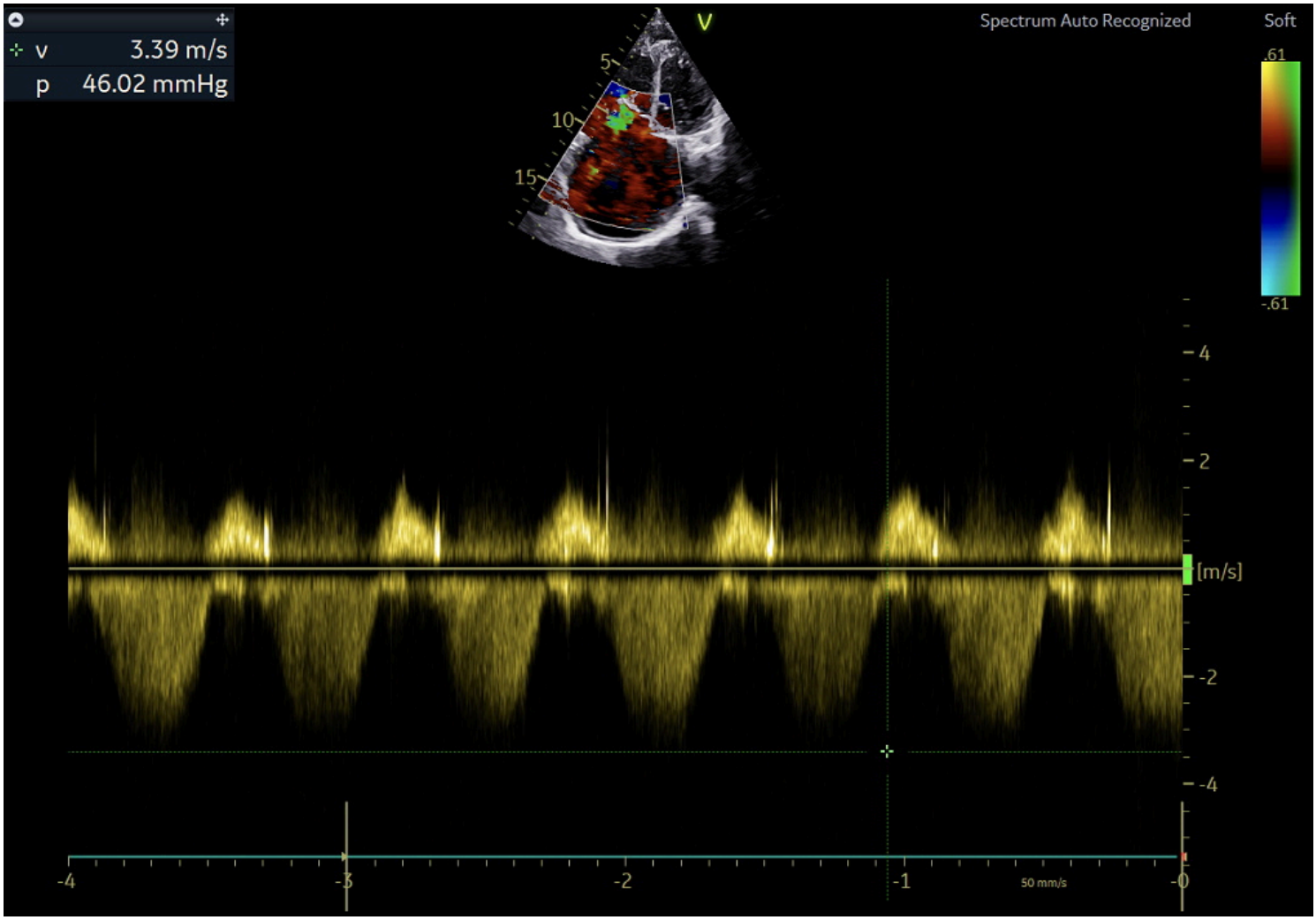

Figure 4

Transthoracic echocardiography (apical four-chamber view) showing a tricuspid regurgitation jet with an estimated right ventricular systolic pressure of **46 mmHg**, excluding right atrial pressure.

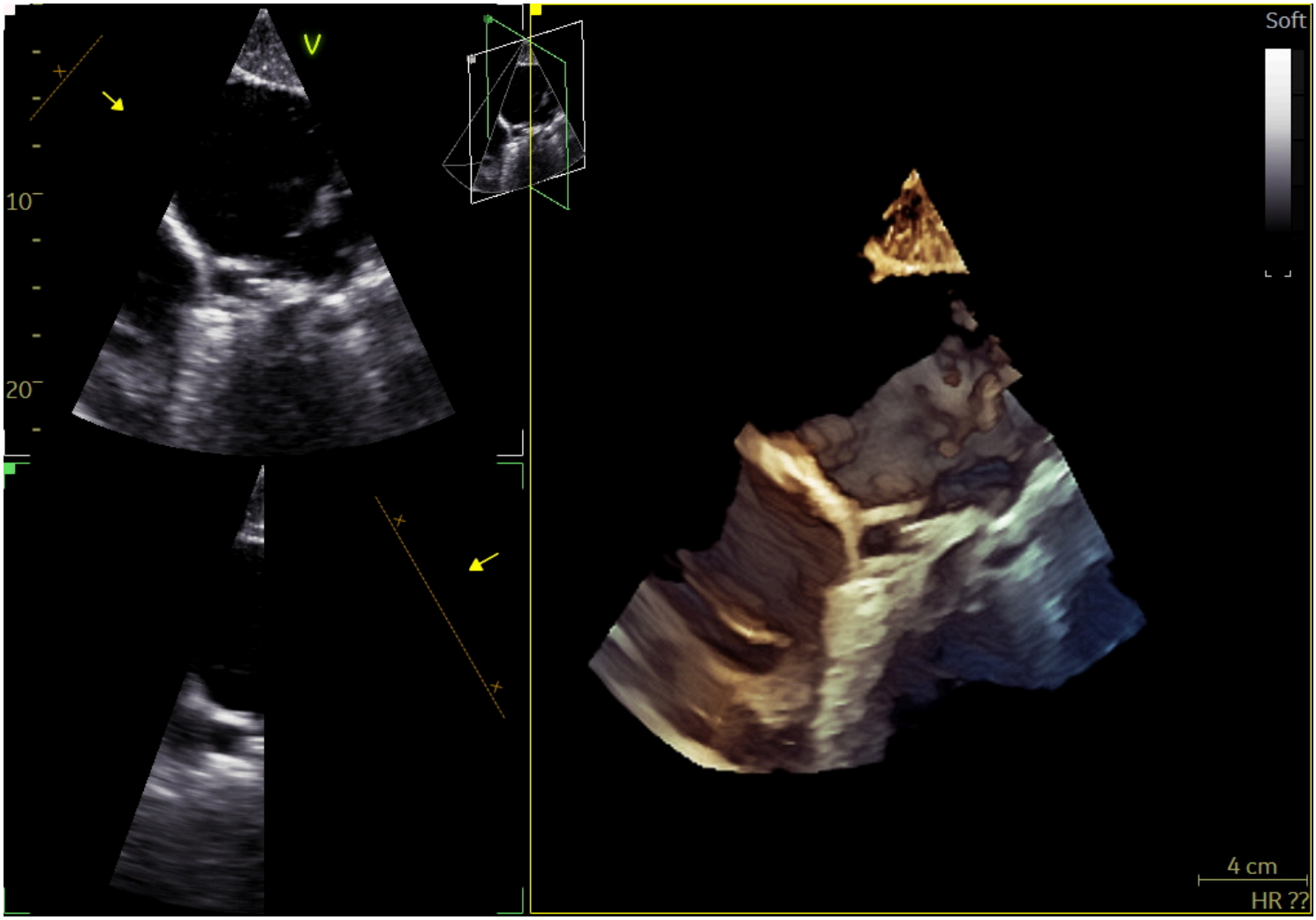

Figure 5

Four-dimensional transthoracic echocardiographic view from the right atrial aspect demonstrating giant right atrial dilatation.

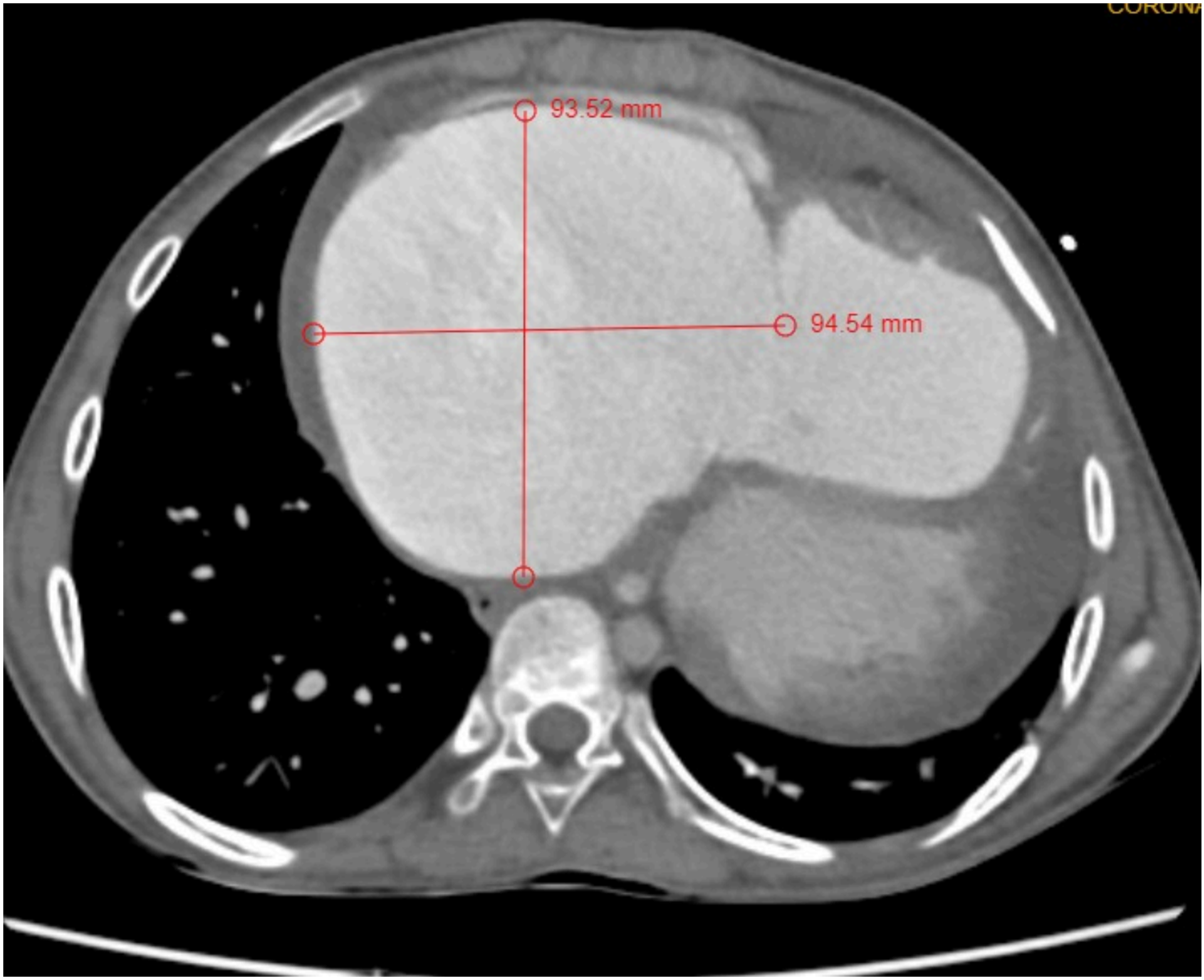

Figure 6

Cardiac computed tomographydemonstrating giant aneurysmal enlargement of the right atrium measuring **9.3 × 9.4 cm**.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- Video01.mov
- Video02.mov