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CASE REPORT

INTERMEDIATE

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE

Acute Left Ventricular Systolic Dysfunction Following Device Closure of Ruptured Sinus of Valsalva Aneurysm

Barun Kumar, DM,[a] Ashwin Kodliwadmath, DM,[a] Pranay Gore, MD,[a] Gaurav Chauhan, MD,[a] Amar Nath Upadhyay, DM,[b] Anshuman Darbari, MCH,[c] Anupam Singh, MS,[d] Ajay Kumar, DM,[e] Nanda N, MD[f]

ABSTRACT

Acute cardiorespiratory distress following device closure of ruptured sinus of Valsava (RSOV) aneurysm can have a battery of differentials. We report a case of acute left ventricular dysfunction with aspiration that caused cardiorespiratory distress immediately following RSOV device closure. Supportive medical therapy led to complete recovery. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:121-127) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

CASE DESCRIPTION

We report the case of a 28-year-old woman who presented to us with an 8-month history of slowly progressive dyspnea and palpitations, preceded by an episode of chest pain, and who was currently in New York Heart Association functional class III, with a history of pedal edema for 2 months. Significant findings on physical examination were a bounding pulse, with a rate of 100 beats/min, blood pressure of 110/38 mm Hg, bilateral pitting pedal edema, jugular venous pressure of 12 cmH_2O with prominent c-v waves, a hyperdynamic apical impulse at the sixth left intercostal space, a grade V/VI continuous murmur best heard at the right sternal border, and an S3. Chest x-ray revealed cardiomegaly with clear lung fields (**Figure 1A**), whereas electrocardiography showed sinus rhythm and low voltage complexes with poor R-wave progression (**Figure 1B**). Transthoracic echocardiography (TTE) showed dilatation and rupture of the noncoronary sinus of Valsalva into

LEARNING OBJECTIVES

- To understand the indications for RSOV closure.
- To decide on the optimal approach for closure of RSOV.
- To be able to make a differential diagnosis of cardiorespiratory distress following device closure of RSOV with a systematic approach.
- To understand the usefulness of multimodality imaging and a multidisciplinary heart team approach in the management of such cases.

From the [a]Department of Cardiology, All India Institute of Medical Sciences (AIIMS), Rishikesh, Uttarakhand, India; [b]Doon Medical College, Dehradun, Uttarakhand, India; [c]Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences (AIIMS), Rishikesh, Uttarakhand, India; [d]Department of Ophthalmology, All India Institute of Medical Sciences (AIIMS), Rishikesh, Uttarakhand, India; [e]Department of Cardiac Anesthesia, All India Institute of Medical Sciences (AIIMS), Rishikesh, Uttarakhand, India; and the [f]Department of Endocrinology, All India Institute of Medical Sciences (AIIMS), Rishikesh, Uttarakhand, India.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

AR = aortic regurgitation

BNP = brain-type natriuretic peptide

CRP = C-reactive protein

LV = left ventricle

LVEF = left ventricular ejection fraction

NCC = noncoronary cusp

PDA = patent ductus arteriosus

RA = right atrium

RSOV = ruptured sinus of Valsalva aneurysm

RV = right ventricle

TEE = transesophageal echocardiography

TTE = transthoracic echocardiography

the right atrium (RA) (Figure 1C and 1D) and dilatation of all 4 cardiac chambers with a left ventricular ejection fraction (LVEF) of 60%. Transesophageal echocardiography (TEE) confirmed the findings; the size of the RSOV was 11.5 mm at the noncoronary cusp (NCC) end (Figure 1E).

QUESTION 1: WHAT ARE THE INDICATIONS FOR CLOSURE OF RSOV?

ANSWER 1. The classical indications for closure of RSOV into RA/right ventricle (RA/RV) are left-to-right shunt >1.5:1, RV volume overload >1.5 cm/m^2, and a margin of the defect at least 5 mm from the right coronary ostium (1). However, heart failure eventually occurs among patients managed conservatively, resulting in death within 1 year; therefore all RSOVs should be closed (2).

QUESTION 2: HOW TO SELECT PATIENTS FOR PERCUTANEOUS VERSUS SURGICAL RSOV CLOSURE?

ANSWER 2. Surgery carries good immediate and long-term results, but currently, it is restricted to the following situations: 1) patients who have an associated ventricular septal defect, especially when juxtaarterial; 2) significant aortic regurgitation (AR), for which an aortic valve resuspension or aortic valve replacement is needed; or 3) failure of catheter closure caused by a large perforation, or prolonged and/or severe hemolysis after device placement. Catheter closure of RSOV has become safe and feasible and is the treatment of choice when there are no associated lesions that require surgery. Approximately 70% of patients with RSOV are potentially eligible for catheter closure (2). Percutaneous closure obviates the need for sternotomy and cardiopulmonary bypass and avoids a second open heart surgery if recurrence occurs because of an acquired fistula (1).

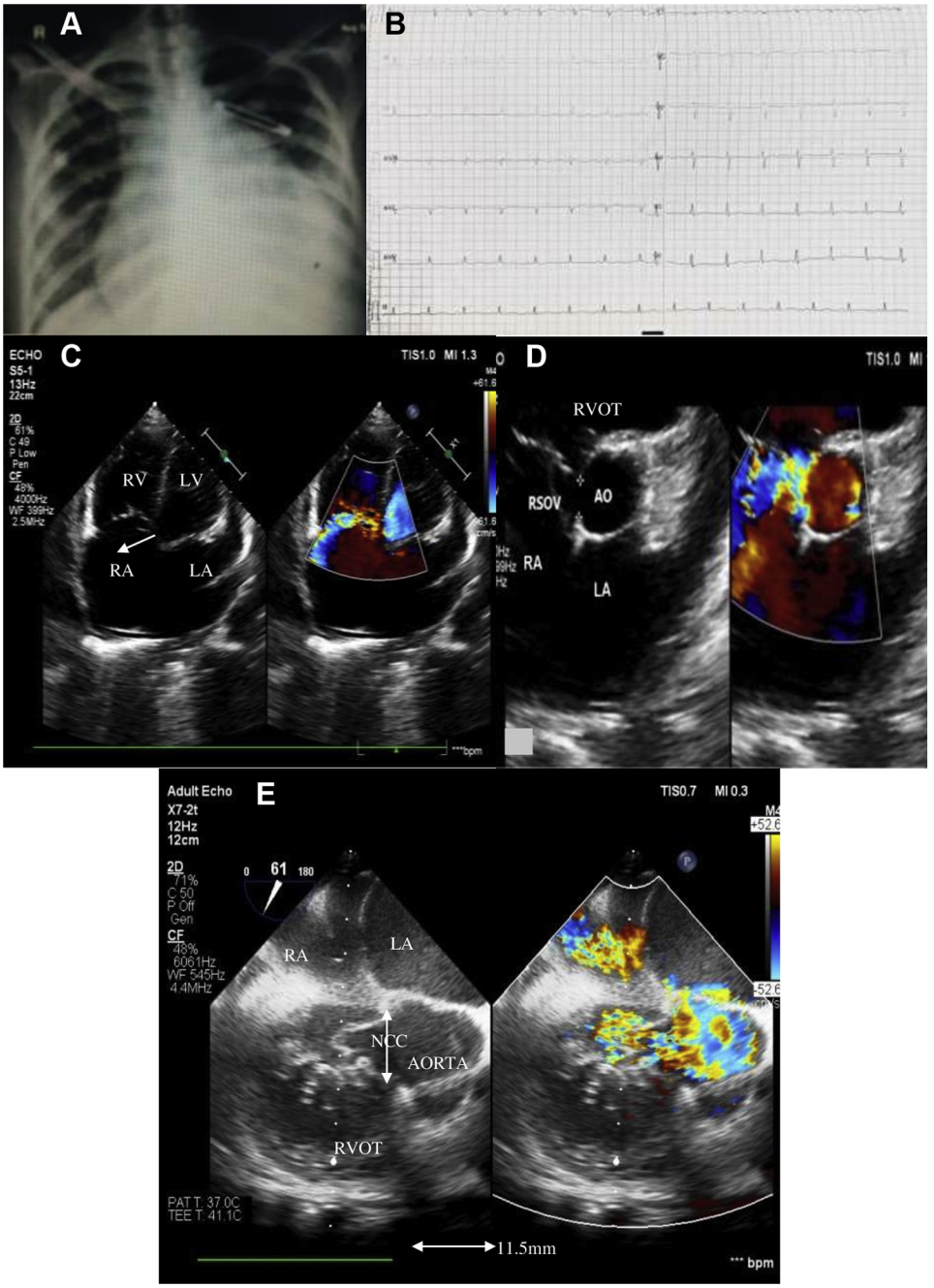
QUESTION 3: WHAT ARE THE STEPS OF PERFORMING PERCUTANEOUS RSOV CLOSURE?

ANSWER 3. After loading with aspirin 300 mg and clopidogrel 300 mg, the patient was taken for device closure of the RSOV using the right femoral route under general anesthesia. After induction of general anesthesia with intravenous propofol 80 mg, fentanyl 80 μg, and vecuronium 10 mg, she was intubated and mechanically ventilated. Unfractionated heparin was used as the anticoagulant during the procedure, and a therapeutic activated clotting time of 250-300 s with a HemoTec (Medtronic ACT Plus Automated Coagulation Timer) machine was maintained throughout the procedure. Aortic root injection showed RSOV draining into the RA, with size at aortic end of 11.8 mm (Figure 2A and 2B). The RSOV was crossed with 5-F MP A1 catheter and 0.035 in × 260 cm straight-tipped Terumo wire from the aortic side into the RA and superior vena cava (Figure 3A). The wire was manipulated and snared through the right femoral vein with a 10 mm snare to form an arteriovenous loop (Figure 3B and 3C). The delivery sheath was passed from the venous end across the RSOV into the aorta (Figure 3D). The size of the device (commonly a patent ductus arteriosus [PDA] duct occluder) should be 2-4 mm larger than the landing zone, which is usually at the aortic end of the RSOV (2). The PDA duct occluder (LT-PDA-1416 [Cera PDA Occluder], Lifetech Scientific Co. Ltd.) was loaded into the sheath, the aortic retention disc was opened into the ascending aorta, and the entire system was pulled back until it was anchored at the aortic end of RSOV (Figure 3E). The other end of device was delivered by stabilizing the loading cable and pulling back the sheath (Figure 3F). After fluoroscopic and TEE confirmation of the proper position and absence of residual shunt and significant AR (Figure 3G), the device was released (Figure 3H). She was extubated and shifted to the coronary care unit with a heart rate of 110 beats/min and a blood pressure of 180/110 mm Hg. Fifteen minutes after shifting to the coronary care unit, she reported worsening respiratory distress. Arterial blood gases showed a partial arterial pressure of oxygen of 66 mm Hg with a saturation of arterial oxygen of 90%, with bilateral coarse crepitations on auscultation.

QUESTION 4: WHAT ARE THE DIFFERENTIAL DIAGNOSES YOU WOULD CONSIDER FOR THE CARDIORESPIRATORY DISTRESS?

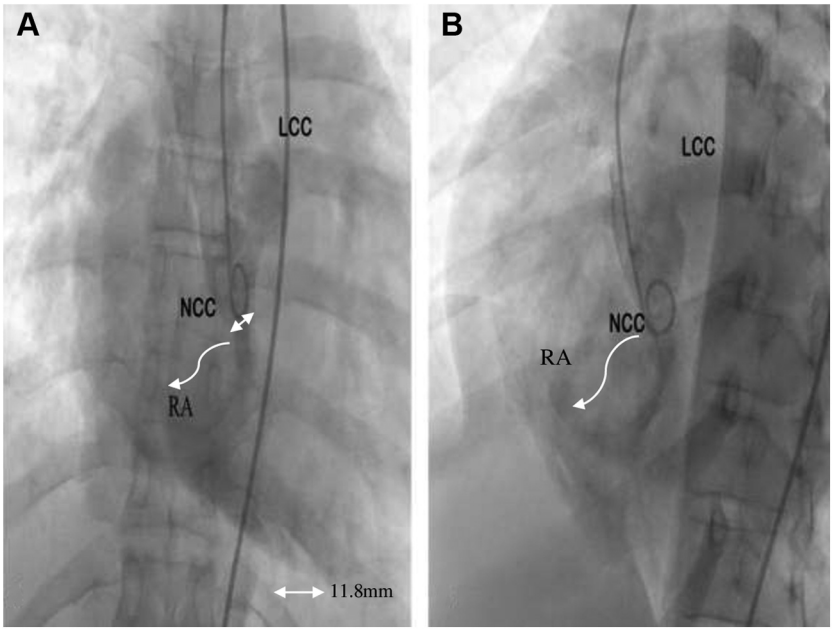
ANSWER 4. Device thrombosis, acute pulmonary thromboembolism, device embolization, acute AR, acute heart failure, cardiac perforation causing pericardial effusion with tamponade, coronary embolism, conduction abnormalities (eg, complete atrioventricular block), pneumothorax, and aspiration were the differentials that needed to be ruled out. Because the RSOV arose from the NCC, and the device was away from the coronaries, the possibility of encroachment of the device on the coronary arteries was negligent (Figure 3H, Video 1).

FIGURE 1 **Preprocedural Chest X-Ray, Electrocardiogram, and Echocardiography**



(A) Chest x-ray bedside anteroposterior (AP) view showing cardiomegaly with clear lung fields. **(B)** Electrocardiogram showing sinus rhythm and low voltage complexes with poor R-wave progression. **(C)** Transthoracic echocardiography (TTE) in apical 4-chamber view showing ruptured sinus of Valsalva aneurysm (RSOV) into right atrium (RA). **(D)** TTE in short-axis view at the level of aortic valve showing RSOV from noncoronary cusp (NCC) draining into the RA. **(E)** Transesophageal echocardiography in short-axis view showing RSOV from NCC draining into the RA. Ao = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle; RVOT = right ventricular outflow tract.

FIGURE 2 Preprocedural Aortogram



(A) Fluoroscopic image in posteroanterior view with pigtail catheter in NCC showing RSOV draining into the RA **(arrow)**. **(B)** Fluoroscopic image in left anterior oblique (LAO) view with pigtail catheter in NCC showing RSOV draining into the RA **(arrow)**. LCC = left coronary cusp; other abbreviations as in Figure 1.

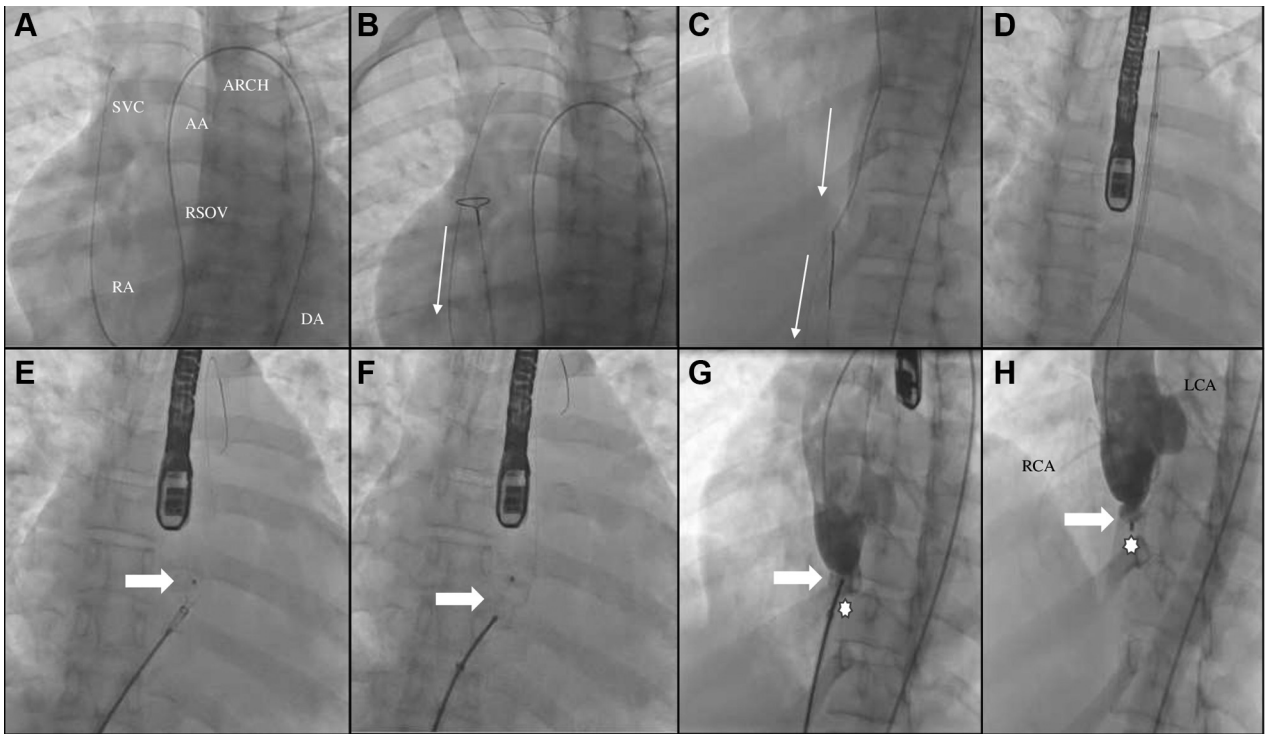
QUESTION 5: WHAT INVESTIGATIONS WOULD YOU CONSIDER AT THIS TIME?

ANSWER 5. Echocardiography confirmed the proper position of the device with no residual shunt, no evidence of device thrombosis or significant AR, but revealed global LV hypokinesia with an LVEF of 10%-15% with moderate mitral regurgitation and mild circumferential pericardial effusion, probably secondary to acute heart failure with no tamponade (Videos 2, 3, 4, and 5). Bedside chest x-ray showed bilateral middle to lower zone (right more than left) opacites in lung fields (**Figure 4A**). An ultrasound of the chest confirmed aspiration (**Figure 4B**). A 12-lead electrocardiogram showed sinus tachycardia and no evidence of a new conduction abnormality. Because the RSOV arose from the NCC, and the device was away from the coronaries (as seen from the aortic root angiogram), and a therapeutic activated clotting time during the procedure decreased the likelihood that a coronary embolism was a cause of the acute drop in EF, a selective coronary angiogram was not performed.

QUESTION 6: HOW WOULD YOU MANAGE THIS CASE?

ANSWER 6. The cardiac anesthesia team reintubated her and connected her to ventilator. She was started on injection furosemide 60 mg bolus, followed by 20 mg every 6 h, nitroglycerin infusion at 5 μg/min, dobutamine infusion at 5 μg/kg/min, dexamethasone 8 mg intravenously every 8 h, dual antiplatelets, and a broad spectrum antibiotic coverage for aspiration. There was no mechanical circulatory support device available. The investigation panel revealed a BNP of 600 ng/L (normal <100 ng/L) and C-reactive protein (CRP) of 160 mg/L (normal <10 mg/L), whereas the renal, liver function tests, complete blood counts, procalcitonin, and cardiac biomarkers were in the normal range. She was extubated the next day after a successful weaning trial and kept on oxygen support. She maintained stable vitals, whereas her echocardiogram still revealed severe LV systolic dysfunction. She was continued on supportive medical therapy along with chest physiotherapy. Chest x-ray on the second postprocedure day

FIGURE 3 Stepwise Percutaneous RSOV Closure



(A) Fluoroscopic image showing MP A1 catheter with Terumo wire being passed from descending aorta (DA) to arch to ascending aorta (AA), across the RSOV to the RA to superior vena cava (SVC). **(B and C)** Snare being passed from femoral vein to snare the MP A1 catheter into the femoral vein to form an arteriovenous loop. **(D)** The delivery sheath being passed from venous end across the RSOV into the aorta. **(E)** Aortic disc of the closure device being deployed **(arrow)**. **(F)** Both discs of the closure device deployed **(arrow)**. **(G)** Aortogram in LAO view showing device across NCC **(arrow)** and no residual shunt **(asterisk)**. **(H)** Aortogram in LAO view after device release showing device across NCC **(arrow)** and no residual shunt (asterisk). LCA = left coronary artery; RCA = right coronary artery; RSOV = ruptured sinus of Valsalva; other abbreviations as in Figures 1 and 2.

revealed improvement in lung haziness (Figure 4C). She improved symptomatically, and oxygen support was removed on the third postprocedure day. On the fifth postprocedure day, TTE revealed an LVEF of 25%-30% with mild mitral regurgitation (Videos 6 and 7). She had no fever during the course of stay, and by day 8, the CRP and BNP levels normalized, and the chest x-ray showed no haziness (Figure 4D). TTE on day 10 showed good recovery of LV function with a LVEF of 50% (Video 8). She was discharged on oral medications, aspirin 75 mg once daily, clopidogrel 75 mg once daily, metoprolol succinate 50 mg once daily, ramipril 2.5 mg once daily, torsemide 10 mg once daily, and spironolactone 25 mg once daily. Follow-up after 1 week and 1 month revealed excellent patient-reported and physician-assessed outcomes, with echocardiography showing normal LV systolic function with LVEF of 55%, with the device in proper position and no residual shunt.

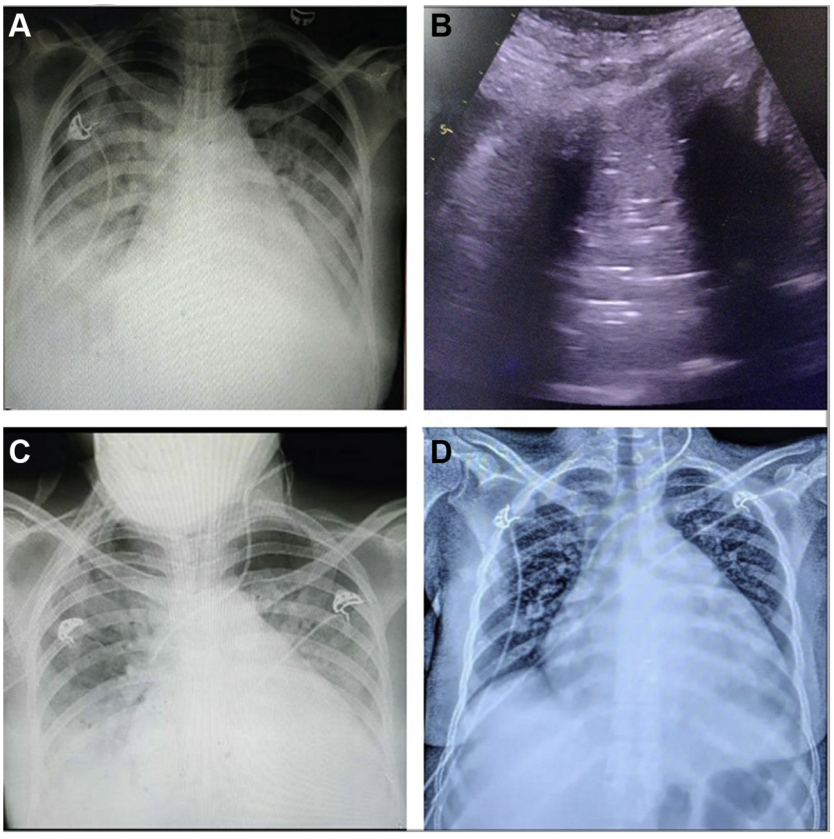
QUESTION 7: WHAT ARE THE COMPLICATIONS OF RSOV DEVICE CLOSURE?

ANSWER 7. Complications include general complications, such as infection, access-site complications, thromboembolic events, and internal bleeding. Specific complications include device migration, hemolysis, encroachment of aortic valve leaflets resulting in AR, atrioventricular conduction disturbances, and right coronary compromise (2).

QUESTION 8: WHAT COULD BE POTENTIAL MECHANISMS OF ACUTE LV DYSFUNCTION IN THIS CASE?

ANSWER 8. Combined use of propofol and fentanyl has been shown to cause acute LV systolic dysfunction with acute heart failure and cardiogenic shock (3). Although the classic triggers for Takotsubo

FIGURE 4 Evolution of Postprocedural Chest X-Ray and Ultrasound Chest Changes Over the Course of Hospital Stay



(A) Chest x-ray AP view bedside done immediately postprocedure showing cardiomegaly with bilateral haziness on the right more than left. **(B)** Ultrasound chest done immediately postprocedure showing minimal pleural effusion, consolidation with aspiration. **(C)** Chest x-ray AP view bedside on second postprocedure day showing decrease in right sided lung haziness. **(D)** Chest x-ray AP view bedside on eighth postprocedure day showing resolution of lung haziness. Abbreviation as in Figure 1.

cardiomyopathy include emotional and physical stress, drug-induced triggers, and other miscellaneous factors (4), it can occur secondary to acute respiratory failure (5) or acute respiratory stress from extubation (6). Although regional wall motion abnormality with LV dysfunction characterizes the classic Takotsubo cardiomyopathy, an atypical variant that causes global hypokinesia has also been described (7).

Although the exact etiology of acute LV dysfunction in our patient could not be elucidated, we assume that it was probably secondary to Takotsubo cardiomyopathy, combined use of propofol and fentanyl, or a combination of the all the previously mentioned factors.

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ADDRESS FOR CORRESPONDENCE: Dr Ashwin Kodliwadmath, Department of Cardiology, All India Institute of Medical Sciences (AIIMS), Virbhadra Road, Rishikesh, Uttarakhand 249203, India. E-mail: ashrocks33@gmail.com. Twitter: @kodliwadmath.

REFERENCES

1. Arora R. Catheter closure of perforated sinus of Valsalva. In: Sievert H, Qureshi SA, Wilson N, Hijazi ZM, eds. *Percutaneous Interventions for Congenital Heart Disease*. 1st ed. Informa Healthcare; 2007:257–262.

2. Kerkar P, DeGiovanni J. Tricuspid valve insufficiency: catheter closure of perforated sinus of Valsalva. In: Sievert H, Qureshi SA, Wilson N, Hijazi ZM, eds. *Interventions in Structural, Valvular and Congenital Heart Disease*. 2nd ed. CRC Press; 2015:427–432.

3. Renilla González A, Lozano Martinez-Luengas I, Eva M Benito Martín, et al. Cardiogenic shock following administration of propofol and fentanyl in a healthy woman: a case report. *J Med Case Rep*. 2011;5:382.

4. Medina de Chazal H, Del Buono MG, Keyser-Marcus L, et al. Stress cardiomyopathy diagnosis and treatment: JACC state-of-the-art review. *J Am Coll Cardiol*. 2018;72(16):1955–1971.

5. Ghadri JR, Bataisou RD, Diekmann J, Lüscher TF, Templin C. First case of atypical takotsubo cardiomyopathy in a bilateral lung-transplanted patient due to acute respiratory failure. *Eur Heart J Acute Cardiovasc Care*. 2015;4(5):482–485.

6. Taniguchi K, Takashima S, Iida R, et al. Takotsubo cardiomyopathy caused by acute respiratory stress from extubation: a case report. *Medicine (Baltimore)*. 2017;96(48):e8946.

7. Ahmad SA, Brito D, Khalid N, Ibrahim MA. StatPearls [Internet]. Takotsubo cardiomyopathy. Accessed January 31, 2021. https://www.ncbi.nlm.nih.gov/books/NBK430798

KEY WORDS cardiorespiratory distress, echocardiography, heart failure, interventional cardiology, percutaneous device closure, ruptured sinus of Valsalva aneurysm

APPENDIX For supplemental videos, please see the online version of this paper.

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