The Journal of Obstetrics and Gynecology of India (July-August 2021) 71(4):440–444
https://doi.org/10.1007/s13224-021-01452-7

CASE REPORT

Successful Outcome of Pregnancy in a Case of Rupture of Sinus of Valsalva Aneurysm: An Interdisciplinary Case Report

T. Ramani Devi[1] · M. Chenniappan[1] · N. Jothi[1]

Received: 20 August 2020 / Accepted: 19 January 2021 / Published online: 26 February 2021

Introduction

Aneurysm of sinus of Valsalva is a rare congenital cardiac anomaly.

This was described in the year 1839 by Hope [1]. It is asymptomatic unless it ruptures causing acute hemodynamic changes. Aneurysm can cause outflow tract obstruction, arrhythmias and heart block.

During pregnancy, increased cardiac output and decreased systemic vascular resistance occur. This can lead to cardiac failure due to intra-cardiac shunts. Altered properties of connective tissue, due to hormones during pregnancy, lead to Rupture of Sinus of Valsalva Aneurysm (RSOVA). This particular case is presented because of its rarity.

Case Report

A 20-year-old primi, married for one year, was referred as a case of cardiac disease complicating pregnancy at 35 weeks of gestation. She was totally asymptomatic. She was diagnosed to have VSD at the age of 1 with no follow-up. During the first trimester, she was detected to have a systolic murmur over the precordium. Initial echo cardiography done outside was interpreted to show a VSD of –6 mm with left to right shunt and mild valvular pulmonary stenosis with normal LV function.

She had regular ante-natal check-up locally and was not on any drugs except for calcium and iron. At 35^{+} weeks, she was referred to our center.

General examination showed normal heart rate, high volume pulse, which was regular and felt equally on all limbs, blood pressure of 120/60 mm Hg, normal SpO_2 and JVP. Cardiovascular examination showed a continuous murmur over the precordium. Her uterine size was 34–36 weeks with good fetal heart.

All investigations were normal. USG showed 34 ± 1 week's fetus with adequate liquor.

2D echo was performed by Cardiologist which showed RSOVA in the right ventricle of size 0.6 cm (Fig. 1), with significant left to right shunt with pressure gradient 154.54 mm Hg (Fig. 2). Left atrium and ventricle were dilated with an adequate LV function. There was also moderate pulmonary stenosis with peak gradient of 44 mm of Hg. There were no aortic regurgitation, no segmental valve motion abnormality, intra-cavitary masses, thrombus, vegetations and pericardial effusion. She was diagnosed as RSOVA into right ventricle and ventricular outflow tract (Fig. 3).

She was advised bed rest and elective LSCS. At 38 weeks, she was admitted with vaginal discharge. Cardiotocography (CTG) did not show any uterine contraction. In view of high risk, elective LSCS was planned. Suddenly, she went in for a precipitate labor. The duration between the onset of contractions and delivery was less than two hours. She was covered with infective endocarditis prophylaxis (ampicillin 2 g I.V. and garamycin 1.5 mg/kg) as soon as she went into labor. Under the supervision

Dr. T. Ramani Devi, MD; DGO; FICS; FICOG is a Consultant Obstetrician & Gynecologist, Ramakrishna Medical Centre LLP, 20, 21, Vivekananda Nagar, Woraiyur, Trichy, 20003, Tamil Nadu, India; Dr. M. Chenniappan, MD; DM; FACC; FACP; FRCP is a Consultant Cardiologist, Ramakrishna Medical Centre LLP, 20, 21, Vivekananda Nagar, Woraiyur, Trichy, 20003, Tamil Nadu, India; Dr. N. Jothi, MD; DA is a Consultant Anesthesiologist, Ramakrishna Medical Centre LLP, 20, 21, Vivekananda Nagar, Woraiyur, Trichy, 20003, Tamil Nadu, India.

✉ T. Ramani Devi
ramanidevidr@yahoo.co.in

M. Chenniappan
chennidr@gmail.com

N. Jothi
drnjothi@gmail.com

[1] Ramakrishna Medical Centre LLP, 20, 21, Vivekananda Nagar, Woraiyur, Trichy, Tamil Nadu 620003, India

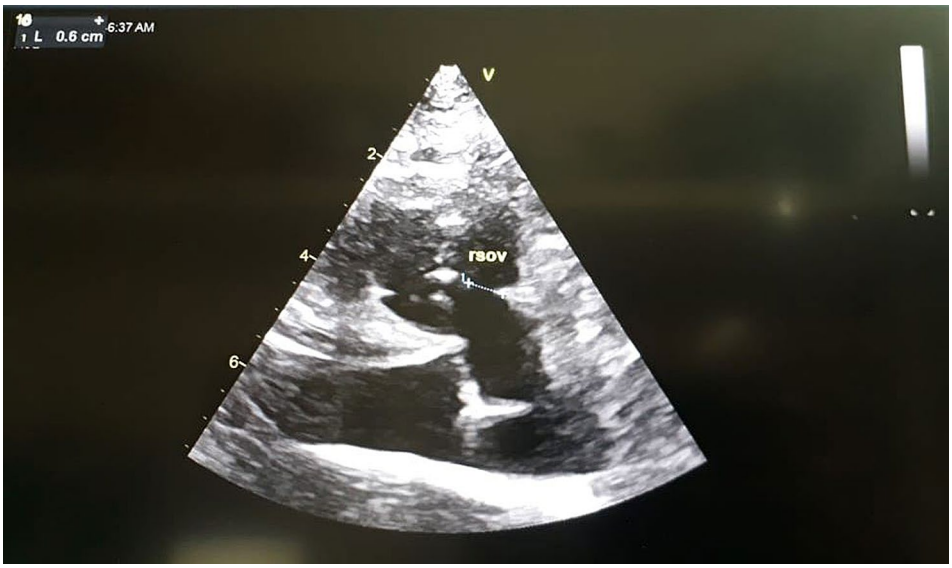

Fig. 1 Echo showing size of the ruptured orifice of RSOVA in RV

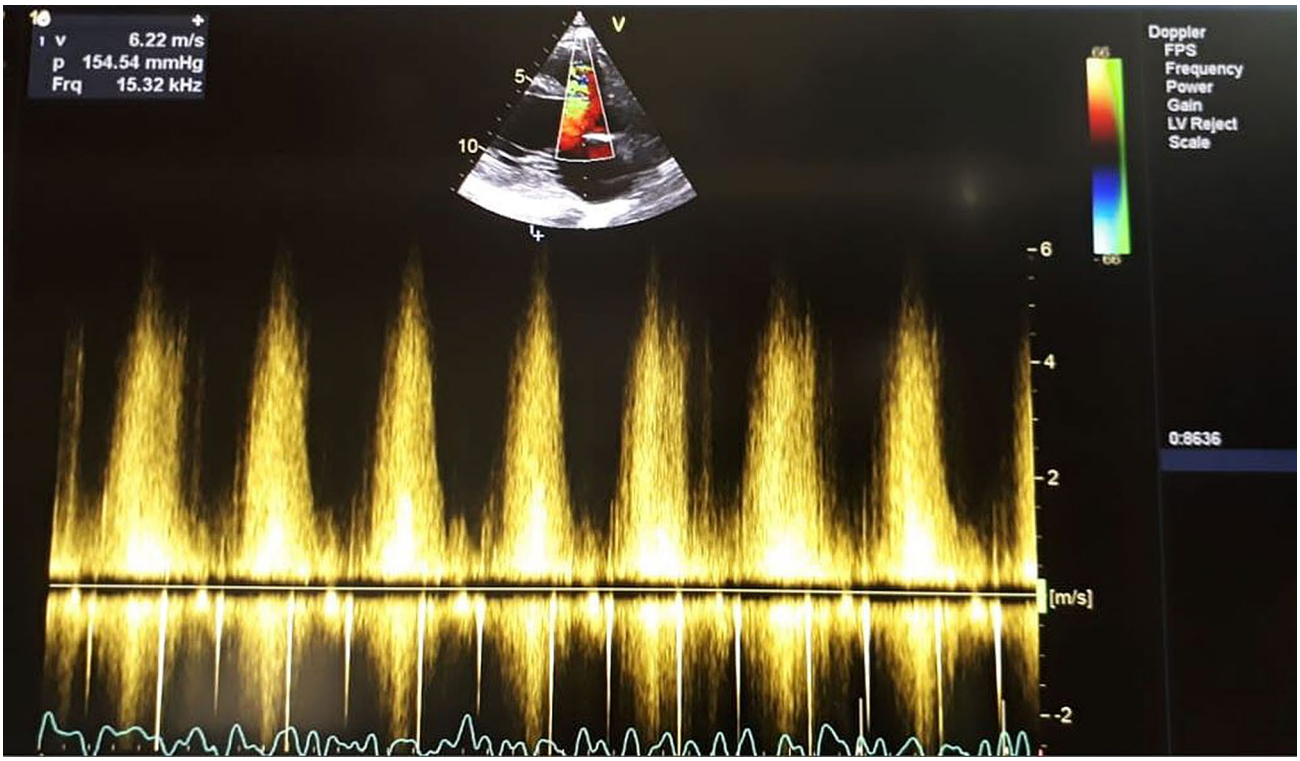

Fig. 2 Doppler showing the peak pressure gradient of RSOVA

of anesthesiologist and cardiologist, she delivered by vacuum (to avoid straining) a female baby weighing 2.8 kg with a good APGAR score. She had mild postpartum hemorrhage which was controlled by uterotonics and observed in the high dependency unit. She was advised contraception and early surgical correction.

Discussion

First case was reported by Thurnam in 1840. It is common among Asians and males [1]. It accounts to 0.1–3.5% of congenital cardiac anomalies and is rarely acquired [1].

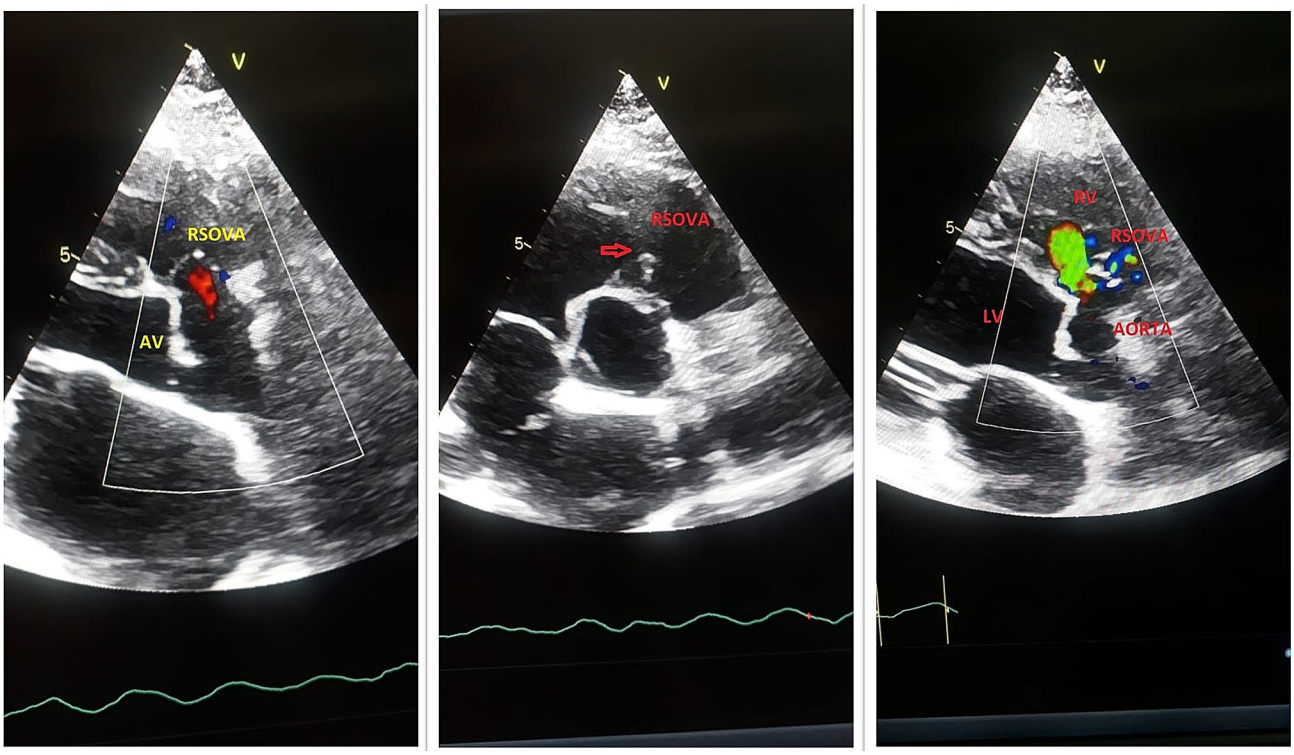

Fig. 3 **a** Short-axis view showing aneurysm, **b** long-axis view showing aneurysm, **c** short-axis view showing left to right shunt into right ventricle due to rupture of right coronary sinus of Valsalva aneurysm

The sinuses of Valsalva are dilatations of aortic valve seen superior to the three aortic wall cusps. They are named as right, left and non-coronary sinuses (Fig. 4). When the tunica media is defective, it leads to aneurysmal dilatation of the sinus of Valsalva resulting in failure of fusion between media and the annulus fibrosis of the aortic valve [2]. 80% patients have aneurysm arising from right coronary sinus, 15.8% from non-coronary sinus and 3.8% from left coronary sinus. 50% of right coronary sinus aneurysm ruptures into the RV leading to VSD, pulmonary valve stenosis and rarely aortic valve regurgitation. Occasionally, this can also rupture cranially into the RA. RSOVA happens when the patient strains unduly [3].

Similarly, non-coronary sinus aneurysms rupture generally into the RA. Left coronary sinus rarely ruptures into the pericardial space leading to cardiac tamponade and

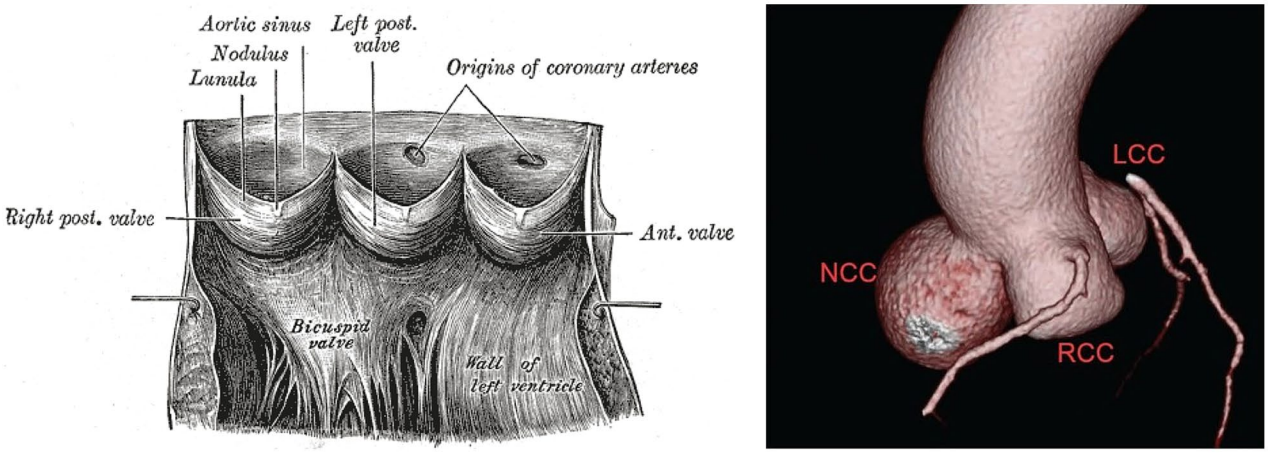

Fig. 4 Normal anatomy of coronary sinuses

imminent death. Sudden death from RSOVA is due to rupture leading to congestive heart failure, cardiac tamponade, arrhythmias or coronary ischemia. Size and location of the shunt determine the prognosis. This anomaly can cause cerebral thromboembolism leading to stroke.

This can also occur in inherited connective tissue disorders like Marfan's syndrome, Ehler Danlos syndrome or inflammatory diseases causing endocarditis, syphilitic or granulomatous aortitis and Behcet's syndrome. It can also arise following stab injuries, aortic dissection or after aortic valve replacement or VSD repair.

Few cases of RSOVA have been reported during pregnancy. At the time of rupture, the patient presents with acute dyspnoea and chest pain; once acute phase is over, in some patients, the symptoms may disappear if the shunt volume is low with restricted RSOVA; otherwise, invariably they land up in heart failure when the shunt volume is large. When left coronary sinus ruptures, it leads to cardiac tamponade. Our patient had chronic rupture; since she had crossed the acute stage earlier, she was asymptomatic when she reported to us. Volume of blood to be shunted from systemic to pulmonary circulation, which leads to right heart failure, depends upon the diameter of rupture.

2D echo and color Doppler (CD) supplemented with 3D reconstruction help in the diagnosis. More than 2D echo and CD, transesophageal echo cardiography (TEE) can clearly map out the lesions. Our patient could not be subjected to TEE because of advanced stage of pregnancy. Computed tomography (CT), magnetic resonance imaging (MRI) and angiography can be supplemental as confirmatory tests in selected cases. Considering the cost and radiation risk of CT, ideal is to go for 2D and TEE. Surgical correction or transcatheter device closure remains the treatment of choice [4].

Ideally, she should be corrected pre-conceptionally. If patient is asymptomatic during pregnancy, we can postpone surgery following delivery. If the patient has congestive heart failure, she can be managed with digoxin and diuretics. Infective endocarditis is also common. In women with significant pulmonary hypertension, there can be right-to-left shunt.

The literature evidence shows RSOVA can be repaired successfully during mid-trimester. In severe symptomatic patients, they can undergo ante-natal cardiac surgery with extra-corporeal support. Now, transcatheter devices have replaced open-heart surgery. Cardio-pulmonary by-pass can lead to placental hypoperfusion and hypoxia leading to fetal death. Fetal mortality can be 19–33%. Gaurav Agrawal et al. have reported a case of RSOVA during pregnancy which was closed by device method, reducing the maternal and fetal risk. Arora et al. have reported 8 patients who were managed with device closure. Abdomen has to be shielded with lead to reduce the radiation exposure. Fetal risk increases with radiation above 50 mGy. Though there is no increased risk of abortion or fetal malformation, still risk of childhood malignancy cannot be excluded.

Goel et al. and Latzman et al. have discussed about the mode of delivery without much complications. Romero et al. have reported a case where LSCS was done under regional analgesia. The literature evidence shows management with LSCS should be taken care similar to other high-risk cases like Eisenmenger syndrome.

Anesthesia for Patients Who are Undergoing LSCS

Patients have to be monitored through ECG, pulse oximetry and CVP. $EtCO_2$ should be maintained around 36–40 mm of Hg and CVP around 3–5 cm of water. LSCS should be planned under epidural analgesia (EA) or general anesthesia (GA). Patient should be positioned to remove the aorto caval compression. EA is ideal, as it reduces the systemic vascular resistance which in turn reduces shunt fraction.

In case of GA, patient should be pre-oxygenated and induced with thiopentone, succinyl choline and lidocaine for stress attenuation. Patient should be intubated with 50:50 of oxygen and nitrous oxide along with isoflurane, vecuronium and fentanyl. After delivery, isoflurane level should be reduced to 1/3rd MAC value. Patient can be reverted with neostigmine and glycopyrrolate. GA is ideal as it has controlled hemodynamic stability. Postoperative analgesia should be maintained [1].

Conclusion

With early diagnosis and proper management in a high-risk pregnancy unit, these patients can have a favorable materno-fetal outcome. Surgical correction is preferable in non-pregnant status. Though LSCS is ideal, vaginal delivery is possible under monitoring.

Compliance with ethical standards

Conflict of interest There are no conflicts of interest to declare.

References

1. Marroush TS, Boshara AR, Botros B, Vendittelli P, Ahmed Z, Dawood L, Rosman HS. Rupture of sinus of Valsalva aneurysm: two case reports and a concise review of the literature. Heart Lung. 2018;47(2):131–5.
2. Fang G, Li M, Li J, Lin L, Mei W. Anesthetic management of cesarean delivery in parturients with ruptured sinus of Valsalva

aneurysm: CARE-compliant 2 case reports and literature review. Medicine. 2017;96(19):e6833.
3. Charfeddine S, Abid D, Triki F, Abid L, Kammoun S, Frikha I. Unusual case of ruptured sinus of valsalva aneurysm in a pregnant woman. Pan Afr Med J. 2017;27:271.
4. Galeczka M, Glowacki J, Yashchuk N, Ditkivskyy I, Rojczyk D, Knop M, Smerdzinski S, Cherpak B, Szkutnik M, Bialkowski J, Fiszer R. Medium-and long-term follow-up of transcatheter closure of ruptured sinus of Valsalva aneurysm in Central Europe population. J Cardiol. 2019;74(4):381–7.

About the Author

Dr. T. Ramani Devi is a Consultant and Managing Director of Ramakrishna Medical Centre LLP and Janani Fertility Centre. She has won 10 gold medals during UG and PG periods. She is a National Vice-President FOGSI 2020 and Chairperson Endometriosis Committee of FOGSI 2014–2016. She has presented in many national and international conferences and authored 16 topics for books and 12 publications in national and international journals. She is a Investigator in POISE trial published in Lancet and edited FOGSI GCPR on endometriosis in 2017. Apart from these, she is the recipient of Dr. A.P.J. Abdul Kalam award 2016, Dr. Navamani Bose and Dr. Rameswary Nallusamy oration, Dr. Dutta's award, Dr. Mehroo Dhara Hansotia award, Dr. S.N. Pandit award and Best Women Doctor Award 2019.