

# Normal Pregnancy and Birth Despite Ruptured Sinus of Valsalva Aneurysm:

Another Case for the Record

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*Sinus of Valsalva aneurysm is a rare cardiac abnormality. Ruptured sinus of Valsalva aneurysms in pregnancy are of course rarer still. We present a case in which an aneurysm ruptured into the right ventricular outflow tract during pregnancy.*

*In 2012, a 26-year-old Chinese woman, in the 18th week of pregnancy and with no apparent evidence of cardiac problems, was diagnosed with atrioventricular septal defects and a sinus of Valsalva aneurysm that had ruptured into her right ventricular outflow tract. After an uncomplicated full-term pregnancy, she gave birth to a healthy baby boy by cesarean section. Fifty days postpartum, the patient underwent surgical repair of the ruptured aneurysm and other cardiac defects. Her surgical outcome was good. As of May 2013, the patient and her baby were healthy.*

*Ruptured sinus of Valsalva aneurysm in pregnancy can be asymptomatic, and women with such a rupture can have a normal full-term pregnancy and give birth to healthy babies. Cesarean section is preferable for pregnant women with ruptured sinus of Valsalva aneurysm because the hemodynamic changes associated with labor can aggravate the aneurysm. Surgical repair should be performed as soon as the patient's condition allows. (Tex Heart Inst J 2016;43(2):158-60)*

**Key words:** Aortic aneurysm/etiology/radiography/surgery; China; congenital heart disease; female; pregnancy complications/diagnosis/surgery; sinus of Valsalva aneurysm/radiography/surgery; vascular fistula

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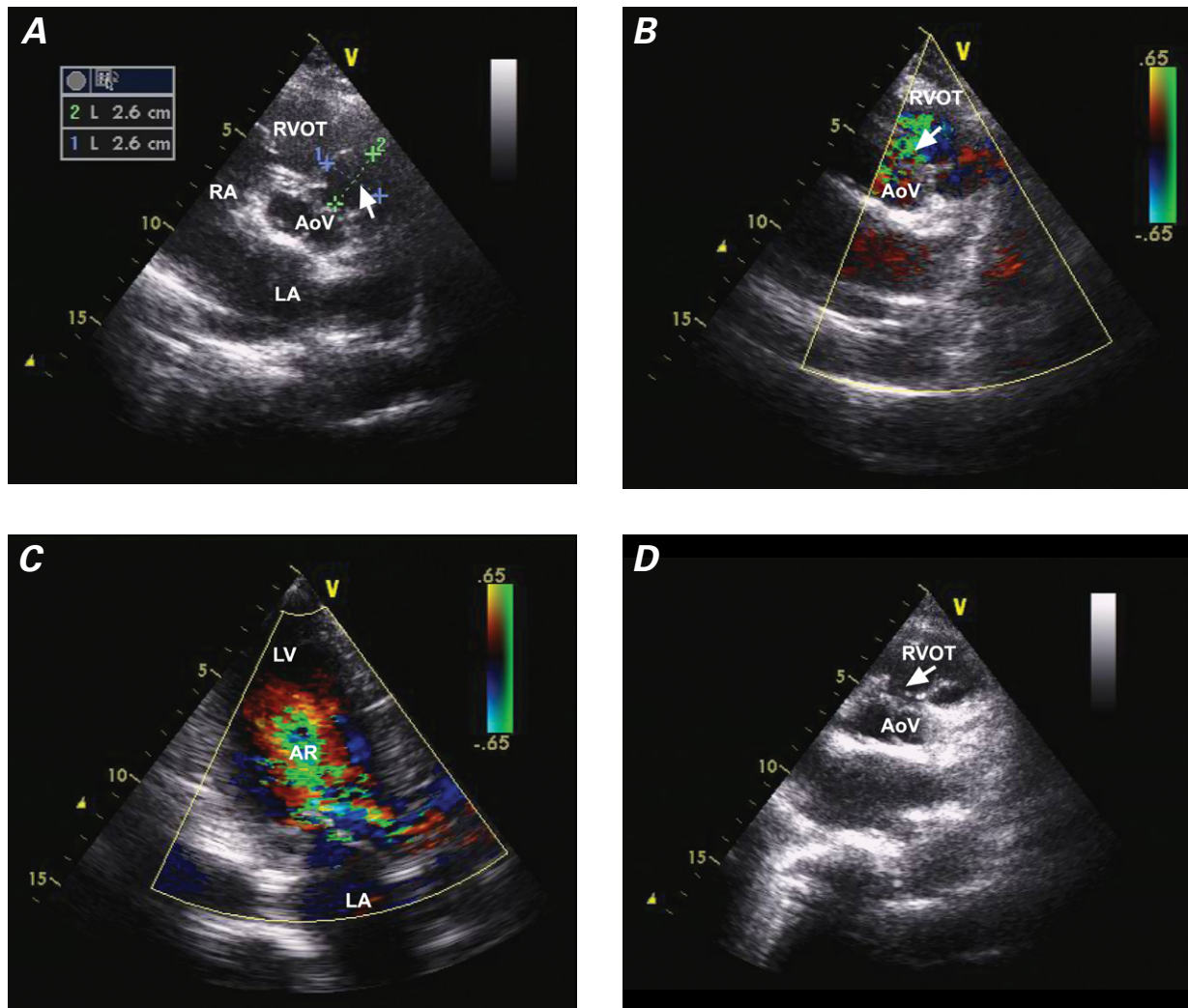
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**S**inus of Valsalva aneurysm (SVA), a rare cardiac abnormality, is either congenital or acquired. The exact prevalence of SVA remains unknown because unruptured SVA is usually asymptomatic. Takach and colleagues<sup>1</sup> reported that the incidence of congenital SVA accounts for 0.1% to 3.5% of all congenital heart defects. Congenital SVA is more prevalent in men than in women and has a particularly high incidence in Asian countries.<sup>2</sup> Approximately 50% of SVAs that rupture into the right ventricle are concomitant with other ventricular septal defects, including pulmonary valve stenosis in most cases and aortic valve regurgitation in some cases. Severely ruptured SVA can lead to heart failure. Therefore, SVA should be treated promptly, regardless of the presence of rupture. Pregnancy combined with ruptured SVA is very infrequent. We report the case of a pregnant patient who presented with a ruptured SVA.

## Case Report

In February of 2012, a 26-year-old Chinese woman was diagnosed at our hospital, in the 18th week of pregnancy, with an SVA that had ruptured into the right ventricular outflow tract (RVOT) and with atrioventricular septal defects (AVSDs). Her physical examination at diagnosis showed a blood pressure of 140/90 mmHg, no apparent evidence of congestion or heart failure, and a grade 4 aortic valve murmur. Transesophageal echocardiography, with and without color-flow Doppler, revealed a significantly enlarged left ventricle (LV) and a right coronary SVA, ruptured and bulging into the RVOT, resulting in acceleration of blood flow in the RVOT (Figs. 1A and B). These echocardiograms also showed severe aortic regurgitation (Fig. 1C) and an approximately 2-cm AVSD below the pulmonary valve (Fig. 1D). Table I shows the diameters of the left atrium, the LV, and the aortic root.

The patient declined our advice to terminate the pregnancy and experienced uncomplicated, full-term gestation. Her heart condition did not progress at the end of



**Fig. 1** Transesophageal echocardiograms during the patient's 18th week of pregnancy show **A**) the right coronary sinus aneurysm (arrow), **B**) the aneurysm's rupture (arrow) into the right ventricle (color-flow Doppler mode), **C**) severe aortic regurgitation (color-flow Doppler mode), and **D**) a ventricular septal defect in the RVOT (arrow).

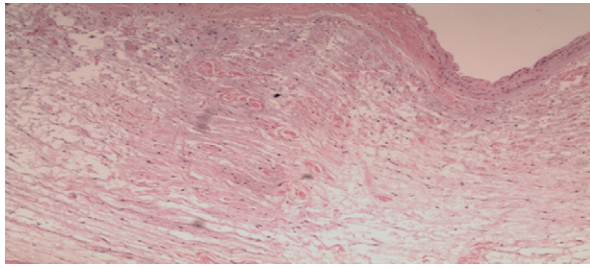
AoV = aortic valve; AR = aortic regurgitation; LA = left atrium; LV = left ventricle; RA = right atrium; RVOT = right ventricular outflow tract

**TABLE I.** Pre- and Postoperative Left Atrial and Ventricular Dimensions

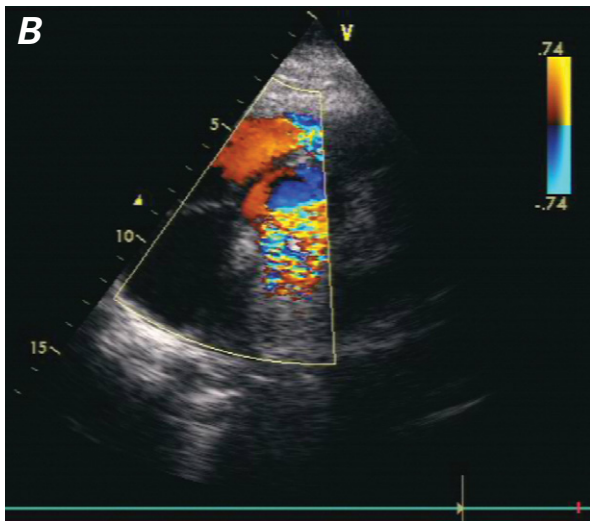
| Time                    | Left Atrium (mm) | Left Ventricle (mm) | Aortic Root Diameter (mm) | Aortic-to-Left Atrial Ratio |
|-------------------------|------------------|---------------------|---------------------------|-----------------------------|
| Preoperative            | 51               | 81                  | 34                        | 0.67                        |
| 7 days postoperatively  | 40               | 56                  | 38                        | 0.95                        |
| 1 month postoperatively | 39               | 54                  | 39                        | 1                           |

the pregnancy. She gave birth to a healthy baby boy by cesarean section and was monitored closely in the intensive care unit afterwards. She had no postpartum complications and was discharged from the hospital 7 days after the operation.

Fifty days after childbirth, the patient, under cardiopulmonary bypass, underwent surgical repair of the AVSD and the right coronary sinus of Valsalva—together with replacement of the aortic valve, the thickened leaflets of which showed evidence of fibrosis and



**Fig. 2** Photomicrograph of a tissue sample from the aortic valve shows fibrosis and myxoid degeneration (H & E, orig. x200).



**Fig. 3** Transesophageal echocardiograms 7 days after cardiac surgery show **A**) the repaired ventricular septal defect and **B**) normal blood flow (color-flow Doppler mode).

myxoid degeneration (Fig. 2). She recovered well from the operation, and the echocardiogram 7 days afterward showed normal heart function (Fig. 3). She was discharged from the hospital 8 days postoperatively. Her normal physical examination at one month enabled her to resume the activities of daily living. As of May 2013, the patient and her baby were healthy.

## Discussion

Ruptured SVA in pregnancy is very seldom reported. In our literature search, we found only 3 cases of ruptured SVA in pregnancy.<sup>3-5</sup> A 22-year-old Turkish Cypriot woman, diagnosed (after presentation with palpitation and chest pain) with SVA that had ruptured into her RVOT in the 37th week of pregnancy, gave birth to a healthy baby by cesarean section; the ruptured SVA was surgically repaired 7 days postpartum.<sup>3</sup> A 26-year-old white American woman, in critical condition from an acute rupture of SVA during early pregnancy, underwent a successful surgical repair of ruptured SVA during pregnancy and delivered a healthy baby vaginally at full term.<sup>4</sup> More recently, Latzman and colleagues<sup>5</sup> reported the case of an asymptomatic 35-year-old Korean woman who presented with a ruptured SVA during pregnancy. The patient had a normal pregnancy and an uncomplicated vaginal delivery, and did not undergo surgical repair for the ruptured SVA after childbirth.

Our patient was asymptomatic during pregnancy and gave birth to a healthy baby by cesarean section. We think that the optimal time for surgical repair of ruptured SVA for pregnant patients should be determined according to the specific condition of the individual patient. If a patient is critically ill or shows obvious symptoms associated with ruptured SVA, surgical intervention should be immediate. To avoid the risk of fetal loss, transcatheter closure of a ruptured SVA might be a better option for pregnant women than is conventional open-heart surgery.<sup>6,7</sup> Even in asymptomatic pregnant patients, fetal safety should be a central factor in determining the optimal time for surgical repair.

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