

## The 4<sup>th</sup> International Congress on Cardiac Problems in Pregnancy (CPP2016)

# RUPTURED SINUS OF VALSALVA (ROSV): A RARE CAUSE OF ACUTE HEART FAILURE IN PREGNANCY

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#### **BACKGROUND**



Rare congenital cardiac defect – 0.1 – 3.5% - deficiency of muscular and elastic tissue in the aortic wall



More common: Asians; males (3-4:1)



Rupture is more common in 20-40y



Associated lesions: VSD, bicuspid aortic valve, aortic coarctation, infundibular pulmonary stenosis



High mortality if surgery is not performed



Few documented cases in pregnancy

#### **CASE REPORT**



lady sought emergency care during her 28<sup>th</sup> week of pregnancy due to sudden shortness of breath and chest pain.



D dimer and venous ultrasound (US) for DVT with no significant alterations



normal left and right ventricular size and function, non stenotic non regurgitant bicuspid aortic valve and a turbulent flow with a not so well defined path that was taken for a small VSD



She was sent home but returned a couple of days later with the same complaint. The same exams were repeated and again, no conclusion was drawn

#### PAST MEDICAL HISTORY

one uneventful vaginal delivery 4 years ago.

regular exercises - 2-3 times a week

a heart murmur 3 years ago

## **CASE REPORT**



A loud continuous murmur, best auscultated over the lower left sternal border



BP 110/60mmHg and HR 92bpm



**Normal breathing sounds** 



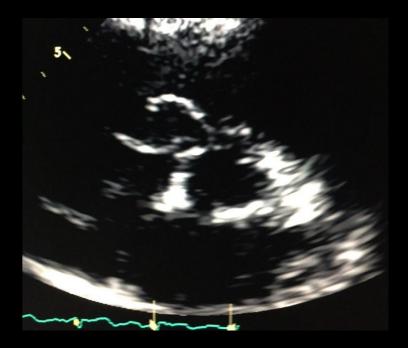
**ROSV** was suspected

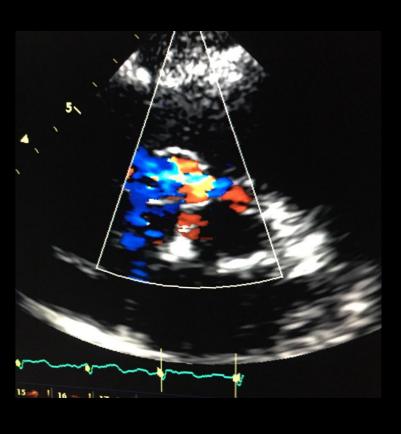


Lab: abnormal NT-proBNP



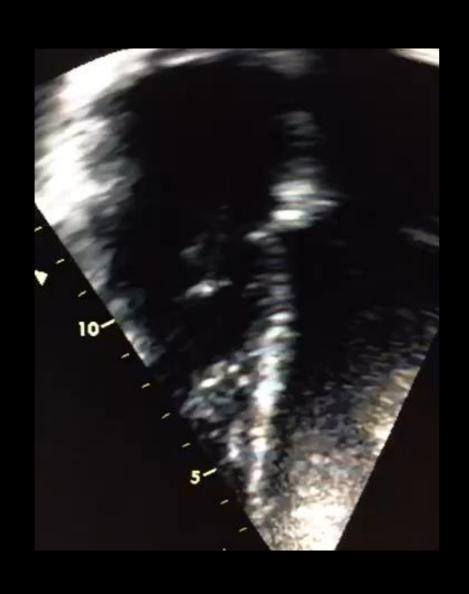
ECHO: repeated in a reference center and confirmed the rupture of the sinus of Valsalva to the right atrium with a large left to right shunt





## **ECHO**





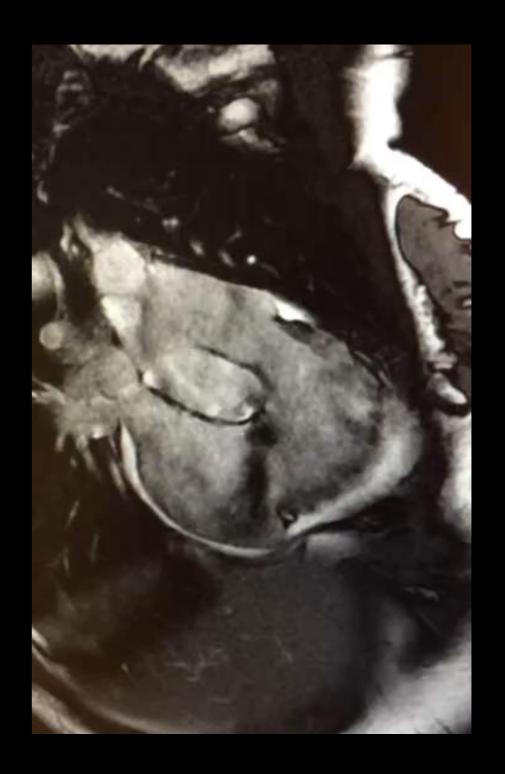


## CASE REPORT - FOLLOW UP

- Mild to moderate symptoms of heart failure persisted in spite of medical treatment (furosemide)
- C-section: on 37th week of pregnancy under combined spinal and epidural anesthesia
- Delivery: no complications
- Postpartum: Heart failure symptoms got worse
- CMR: confirmed the bicuspid aortic valve and "windsock" RSOV in the RA
- Percutaneous treatment 20 days postpartum
- Two month follow up: patient was asymptomatic with no medications or residual shunt by ECHO.

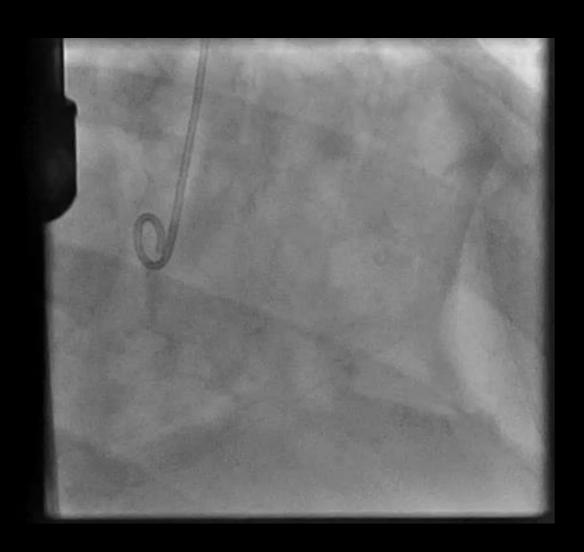


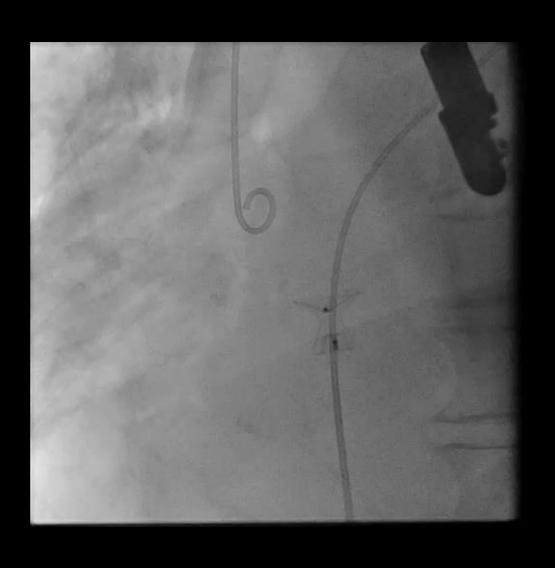






# Transcatheter closure of rupture of sinus valsava aneurisms with Amplatzer Occluder





## **Discussion - ROSV**

Congenital

Inherited connective tissue abnormality (Marfan, Ehlers-Danlos)

Trauma / latrogenic

Bacterial endocarditis

Infections:
syphilis,
tuberculosis

## **DIAGNOSIS**

#### SYMPTOMS / SIGNS

Asymptomatic till rupture Acute heart failure (40%)

- Dyspnea
- Chest pain
- Palpitations
- Fatigability

**Heart block** 

Death
 Loud continuous heart
 murmur /thrill
 Aortic regurgitation

#### **ECHOCARDIOGRAM**

- 90% accuracy
- QP/QS 2.3 (1.2-4.3)
- Main error: identification of drainage chamber

#### DISCUSSION: PREGNANCY



Hormone induced changes in aortic wall



Hemodynamic burden during pregnancy



**Impact of Labor:** 

extra load could enlarge the

tear and the shunt



Type and time of delivery:

balance between mother and fetus

### TREATMENT

#### PROGNOSIS IS POOR IF NOT CORRECTED

#### **SURGERY**

- Asymptomatic unruptured
- Progressive enlargement
- Symptomatic

#### **PERCUTANEOUS**

- Less invasive
- Effective in more than 90% of cases
- Contraindicated:
   moderate to severe AR;
   other cardiac diseases
   with indication to open
   heart surgery

## CONCLUSION

This case illustrates the value of clinical signs that allow proper diagnosis and management of rare diseases that could threaten mothers' lives during pregnancy or postpartum.