

# SECUNDUM ATRIAL SEPTAL DEFECTS COMPLICATED WITH SEVERE PULMONARY HYPERTENSION IN PREGNANCY: MANAGEMENT AND OUTCOME

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## ABSTRACT

**Background:** Atrial septal defects (ASD) are the third most common type of congenital heart disease, that allows shunting of blood between the systemic and the pulmonary circulations. ASD complicated with severe pulmonary hypertension (PH) often occurs in median age, and pregnancy should be prevented since this will contributed to both high maternal mortality, and complication in pregnancy.

**Objective:** To discuss management and outcomes of secundum atrial septal defects complicated with severe pulmonary hypertension in pregnancy.

**Case:** A 21-year-old woman, G2P1AO, referred to Maternal Fetal Medicine Division on March 30, 2017 with 31 weeks of pregnancy and shortness of breathing. Further investigation revealed that she had history of atrial septal defects that was diagnosed 2 years before admission. Ultrasound examination was performed with conclusion 31 weeks intrauterine pregnancy with intrauterine growth restriction (IUGR). Echocardiography result indicated large secundum ASD, severe PH, severe tricuspid regurgitation (TR), severe pulmonary regurgitation (PR), and ejection fraction (EF) 43%. After closed discussion with the patient and family, the team decided to perform C-section after lung maturation. On April 10, 2017 patient underwent C-section under epidural analgesia with the pregnancy results female baby, 1170 g, APGAR Score 7/8. The patient treated in ICCU for 3 days and went home after 7 days of treatment.

**Conclusion:** Large secundum ASD complicated with severe PH in pregnancy should be carefully managed. Pregnancy termination should be considered, particularly if the disease threatened the mother and caused IUGR in pregnancy.

**Keywords:** Pregnancy, ASD, PH, management.

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## BACKGROUND

Atrial septal defects (ASD) are the third most common type of congenital heart disease, that allows shunting of blood between the systemic and the pulmonary circulations.<sup>1</sup>

Atrial septal defect (ASD) was first described by Rokitansky in 1875. Its clinical features were described in 1941 by Bedford et al, and became well known with the writings of Barber et al, in the 1950s. After bicuspid aortic valve it is the most common congenital defect in adults. Fuster et al

reported an incidence of 22% among adults with congenital.<sup>2,3</sup>

Atrial septation involves septum primum, septum secundum, and the atrioventricular canal septum. About 65–70% of patients with a secundum defect, roughly 50% of those with a primum atrial septal defect, and 40–50% of those with a sinus venosus defect are female.<sup>3,4</sup>

Secundum atrial septal defect is a defect within the fossa ovalis usually due to one or several defects within septum primum. Most secundum defects are not confluent with the vena cava, right pulmonary veins, coronary sinus, or the atrioventricular valves. ASD complicated with severe pulmonary hypertension (PH) often found in median age, and pregnancy should be prevented due to both high maternal mortality, and complication in pregnancy.<sup>4,5</sup>

**Objective:** To discuss management and outcome of secundum atrial septal defects complicated with severe pulmonary hypertension in pregnancy.

**Case:** A 21-year-old woman, G2P1AO, referred to Fetomaternal Division on March 30, 2017 with 31 weeks of pregnancy and shortness of breathing. Three years before admission her first child was delivered spontaneously, weight 4000 gr. No history of shortness of breathing during her first pregnancy. Two years before admission she start to complain about shortness of breathing, and fatigue. She went to hospital and further more she was diagnosed with atrial septal defects. She starts to take her

medicine regularly, but after she found out that she was pregnant for her second child, she stop taking medication.

In her 30 weeks of pregnancy, she started to develop shortness of breathing, she must sleep with 2-3 pillows under her head, and she can't walk more than 10 meters. Her husband than bring her to the hospital. And because of her complication the patient then referred to Maternal Fetal Medicine Division.

Further physical examination found increased superior vena cava pressure, rales on both lungs. Ultrasound examination was performed with conclusion 31 weeks intrauterine pregnancy with symmetrical intrauterine growth restriction (IUGR).

We consulted the patient to Cardiology Department and the echocardiography was performed with result indicated large secundum ASD, severe PH, severe tricuspid regurgitation (TR), severe pulmonary regurgitation (PR), and ejection fraction (EF) 43%. Patient was given close cardiovascular monitorization, intensive oxygen, oral Sildenafil 2x6,25 mg, Furosemide 1x40 mg, and Spironolacton 1x12,5 mg.

After closed discussion with the patient and family, the team decided to perform C-section after lung maturation. We gave Dexamethasone for lung maturation. On April 10, 2017 patient underwent C-section under epidural analgesia, with the pregnancy results female baby, 1170 g, APGAR Score 7/8. Dobutamine titration was given during C-section, the vital sign remain stable; blood pressure between

110-125/ 60-72 mmHg, heart rate 134x/ minute, respiratory rate 30x/ minute, oxygen saturation 95%. We also performed Pomeroy tubectomy.

After C-section the patient treated in ICCU for 3 days, and went home after 7 days of treatment.

The baby was treated in NICU, but after 10 days of treatment, the baby passed away.

After hospitalization the patient then regularly treated as outpatient in Cardiology Department. And nowadays her condition remains stable, she was able to perform her daily activity with no shortness of breathing, she can sleep with only one pillow. She was advised to performed curative surgical treatment, but due to her low social economy she still refused it.

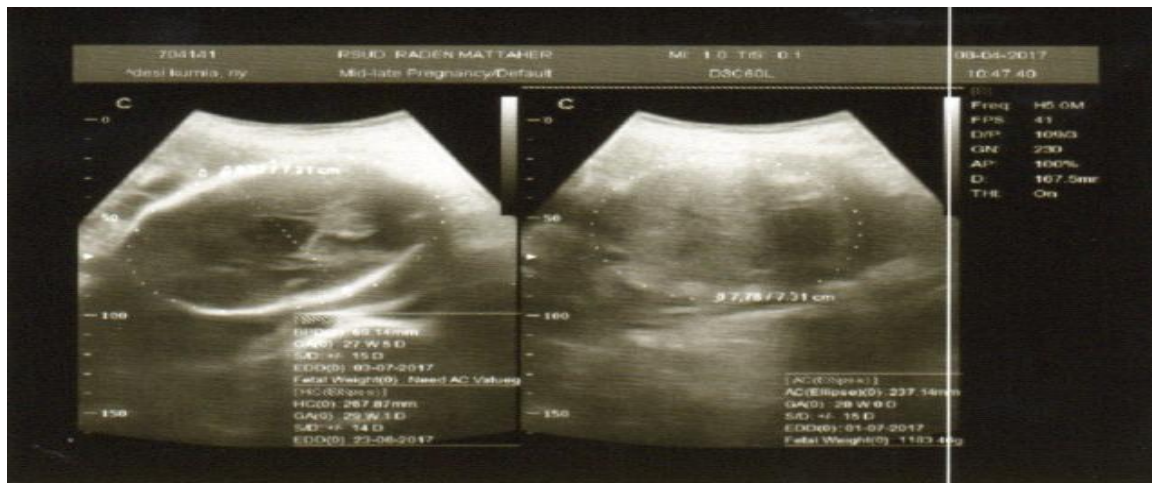


Figure 1. Ultrasound result

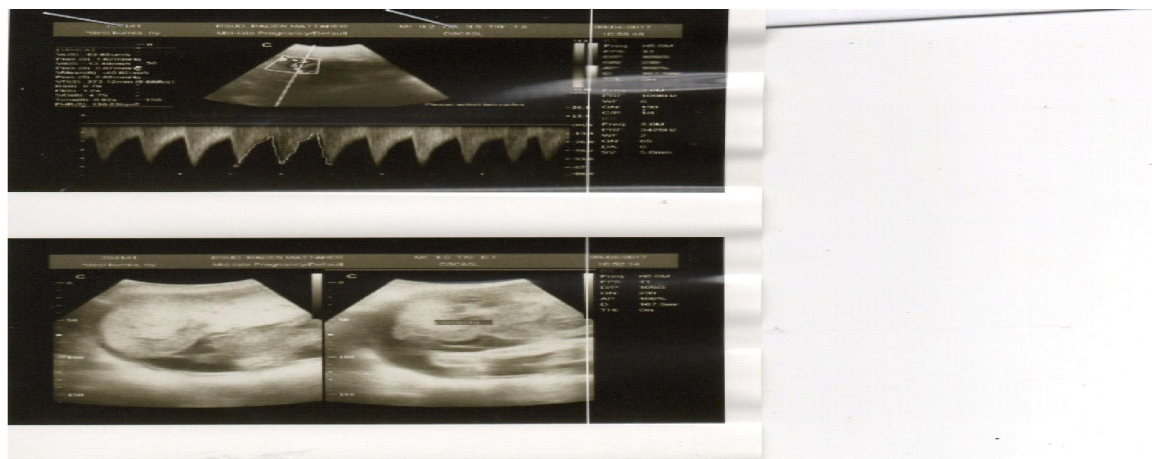
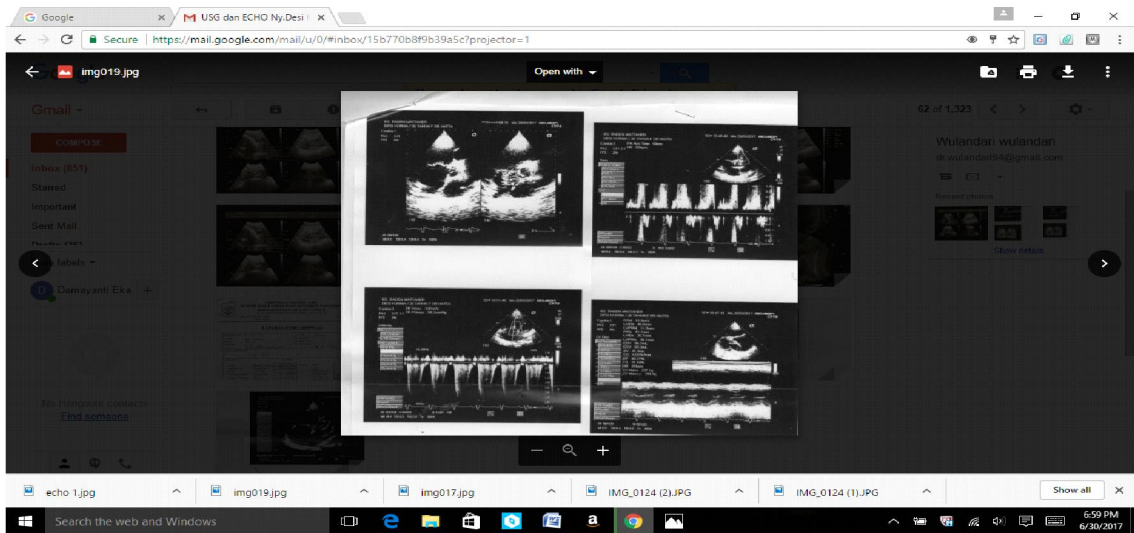
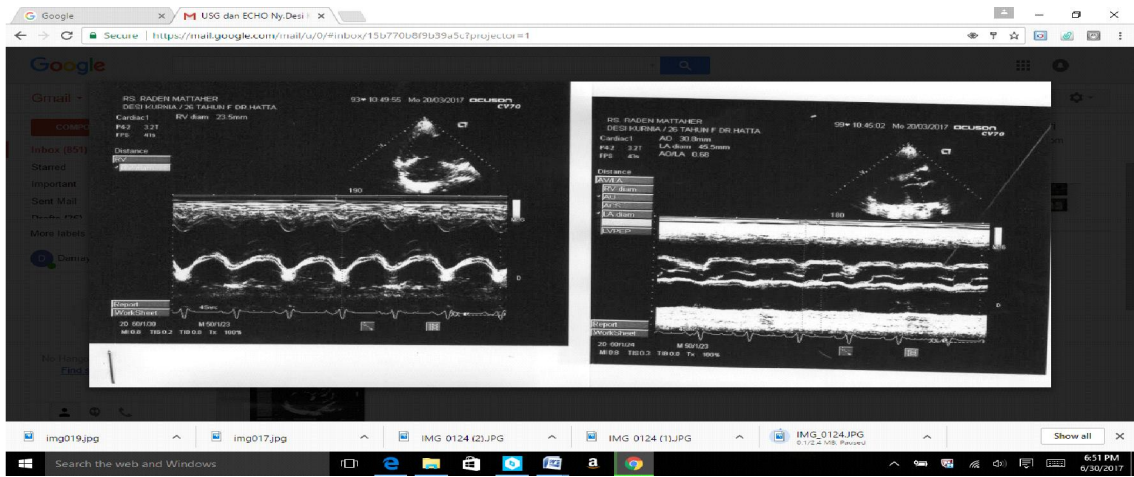


Figure 2. Umbilical artery Doppler





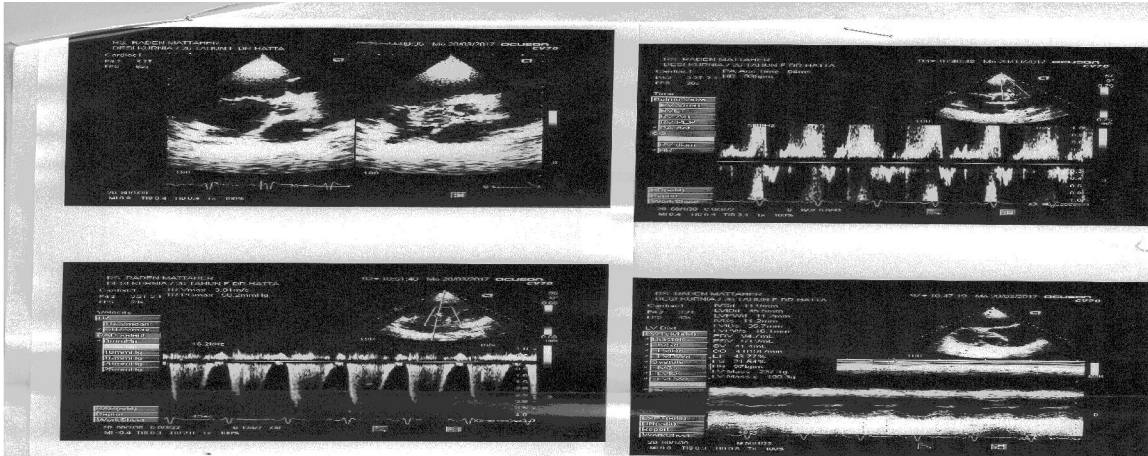


Figure 3. Echocardiography result



Figure 4. Neonatal outcome

**DISCUSSION**

Atrial septal defects belong to a group of congenital cardiac anomalies that allow communication between the left and right sides of the heart.<sup>2,3</sup>

The size of secundum defects varies from several millimetres to 2–3 cm. Large defects are usually associated with substantial deficiency, or even complete absence of septum primum. Most adult patients with a large defect present with symptoms, including fatigue, exercise

intolerance, palpitations, syncope, shortness of breath, peripheral oedema, manifestations of thromboembolism, and cyanosis.<sup>6,7</sup>

A longstanding shunt results in impaired right atrial reservoir and pump functions, right ventricular dilatation, myocardial cell hypertrophy and fibrosis, and cellular injury manifesting as increased serum concentrations of cardiac troponin-I. The pulmonary vascular bed remodels with

myointimal cell proliferation, increased medial smooth muscle, and increased collagen leading to arteriolar narrowing and pulmonary hypertension.<sup>8,9</sup>

In this case we found large secundum ASD, untreated for the past few months and already complicated with pulmonary hypertension, characterized by progressive shortness of breathing, exercise intolerance, and fatigue.

Patient was treated with Sildenafil (PDE-5 Inhibitors). Sildenafil causes vasodilation of the pulmonary vascular bed and in the systemic circulation. It also has a positive inotropic effect on the hypertrophic right ventricle. It is a category B medication. Using sildenafil to treat PH in pregnancy has been described in case reports and appears to be safe, but experience is still limited.<sup>9,10</sup>

Mode of delivery and anesthetic management remain debated. Vaginal delivery may be preferred over cesarean section to minimize postsurgical fluid shifts or increased anesthetic risks. Cesarean section, on the other hand, provides for a more controlled setting, avoids a prolonged second stage of labor, the potential for uncontrolled vaginal hemorrhage, and the adverse hemodynamic effects of bearing down.<sup>11,12</sup>

The goals of anesthetic management are to avoid pain, hypoxemia, hypercapnia, and acidosis; all of which lead to increased PVR and thus hypertension. Spinal and general anesthesia causes peripheral vasodilatation and may worsen the patient's hemodynamic. Regional anesthesia may be advantageous, however, when used in

large dosages, may produce a decrease in venous return because of a sympathetic block.<sup>12,13</sup>

Patients receiving general anesthesia were four times more likely to die, compared with patients receiving regional anesthesia (OR 4.37, 95% CI 1.28–16.5,  $P=0.02$ ). Primigravidae were at higher mortality risk compared with parturient with previous pregnancies (OR 3.70, 95% CI 1.15–12.5,  $P=0.03$ ).<sup>14,15</sup> Therefore in this case the team decided to performed C-section under epidural analgesia. And to avoid future pregnancy, we also performed tubectomy as permanent contraception.

Pregnancy should be avoided in women with an atrial septal defect and severe pulmonary hypertension. In a contemporary study maternal mortality was prohibitively high (28%) in women with congenital heart disease. Maternal deaths tended to occur shortly after delivery and were often caused by heart failure, thromboembolism, pulmonary hypertensive crisis, and sudden cardiac death.<sup>12,13,14</sup>

Yap and colleagues found maternal complications including arrhythmias (4%) and transient ischemic attack (1%). Pre-pregnancy history of arrhythmia and maternal age older than 30 years were risk factors for maternal cardiac complications. By comparison with the general population, women with unrepaired atrial septal defects had an increased risk of pre-eclampsia, fetal loss, and low birth weight.<sup>15</sup>

In this case we found that the maternal complication from ASD including pulmonary hypertension, and IUGR.

A multidisciplinary approach together with pulmonary vasodilator therapy may be successful in such a high-risk pregnant woman.<sup>16</sup>

## CONCLUSION

Large secundum ASD complicated with severe PH in pregnancy should be carefully managed. Pregnancy termination should be

considered, particularly if the disease threatened the mother and caused IUGR in pregnancy.

A multidisciplinary approach together with pulmonary vasodilator therapy, regional analgesia, and cardiovascular monitoring may be successful in such a high-risk pregnant woman.

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