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Editorial

Pregnancy in Patients with Pulmonary Arterial Hypertension "To Deliver, or Not to Deliver: That Is the Question"

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▸ See the article "Pulmonary Arterial Hypertension and Pregnancy: Single Center Experience in Current Era of Targeted Therapy" in volume 49 on page 545.

"To be, or not to be: that is the question."
William Shakespeare, Hamlet, Act III, Scene I

This famous phrase is a soliloquy of Prince Hamlet in the William Shakespeare's play Hamlet. Many PAH patients are young women and they always have the possibility of being pregnant. If we meet a young pregnant woman patient with pulmonary arterial hypertension (PAH), we will have severe mental anguish in the determination of the termination or the maintenance of the pregnancy. We must consider maternal and fetal outcomes in case of pregnancy in PAH patients. Because there are high mortality rates of mother and fetus associated with pregnancy in PAH,[1-3] current treatment guidelines recommend the avoidance of pregnancy in PAH patients clearly and strongly.[4-6] The reported maternal death rate was about 30% to 56% in these previous studies, and the major causes of deaths are right ventricular failure and stroke through intracardiac shunts. Moreover, the fetal death rate was about 11% to 28% usually from premature birth and growth retardation.[3] However, these data resulted from previous studies more than 20 years ago. Thus, we need more recent data reflecting current PAH treatment. In this current issue, Lim et al.[7] reported their data about the clinical outcome of 10 pregnancies in 9 PAH patients. Fortunately, the authors reported there was no maternal or fetal mortality with their multidisciplinary team approach. However, there were severe cardiac events in two mothers despite careful treatment.

All pregnant PAH patients should be counseled to terminate the pregnancy seriously, especially in patients with worsening right heart failure or with other high risk features.[5][6] Therapeutic abortion is recommended before 22 weeks of gestation.[5] Early planned terminations in PAH patients are usually safe.[8] However, if patients want to continue their pregnancies, these patients should be managed by a multidisciplinary team including PAH specialists, cardiologists, anesthesiologists and neonatologists. Because calcium channel blockers, phosphodiesterase type 5 inhibitors, and prostaglandin and its analogues are reported safe during pregnancy,[8-10] current guidelines recommended that pregnant PAH patients should be treated with PAH-specific agents except endothelin-receptor antagonists. In this current study, 90% of them were treated with PAH-specific treatment including sildenafil and treprostinil.[7] These pregnant patients should be closely monitored including monthly follow-up visits in the first and second trimester. They are recommended to have

a weekly clinic visit in their third trimester. The cesarean section with epidural or spinal-epidural anesthesia is the preferred method of delivery. Because the clinical course of PAH can be worsened during the postpartum period, these patients should be monitored intensively.

Editorial members were seriously concerned that this paper might give misconceptions that female patients with PAH may be pregnant safely. However, we decided to accept this paper to give information to readers this valuable experience reflecting current treatment status. Thus, this data should only be applied in PAH patients with unavoidable pregnancy.

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