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Pregnant 30-year-old with idiopathic pulmonary arterial hypertension

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A 30-year-old woman was hospitalized for the first time due to resting dyspnea. At the time of admission she was in the WHO IV class, with lowered arterial blood saturation and echocardiographic features of right ventricle overload. A right heart catheterization (RHC) revealed severe reactive pulmonary arterial hypertension (PAH). Treatment with sildenafil and diltiazem led a reduction of symptoms associated with the WHO II class. The patient was advised to avoid pregnancy. Two months later we noted a significant clinical improvement, but the blood test showed elevated beta-hCG. The risk of pregnancy related complications in PAH patients was discussed, and therapeutic options were proposed, including those related to termination of the pregnancy. The patient's decision was to maintain the treatment and the pregnancy was not terminated.

During the thirty weeks of gestation the patient did not present any symptoms and the course of pregnancy was not complicated. In the 31st week, rapid clinical deterioration occurred. She was urgently admitted to the hospital with severe resting dyspnoea, tachycardia, blood pressure 90/70 mmHg and oxygen saturation of 90–92%. Transthoracic echocardiography (TTE) showed signs of severe right heart pressure overload. We planned for possible escalation of therapy with permanent intravenous epoprostenol infusion, but we had to expect for epoprostenol delivery and due to the patient's critical and life-threatening condition, emergency inhalations of iloprost was administered. RHC was performed trying to avoid excessively exposing the fetus with X-rays. The severe non-reactive PAH was confirmed. On the 3rd day of hospitalization, the patient developed a respiratory tract infection, due to the negative result of the influenza screening test. Eempirical antibiotic therapy was introduced, but the patient's condition continued to deteriorate — leukocytosis and heart rate increased, O2 saturation and blood pressure dropped. Immediately, an urgent specialist consultation was carried out, attended by a cardiologist, cardio-anesthesiologist, neonatologist and gynecologists. The state of the fetus was regarded as a stable, so the decision was made to start an intravenous infusion of epoprostenol and then to perform a Caesarean section. Within three hours of the epoprostenol infusion, the patient had moderate bleeding from the genital tract. Placental ablation or epoprostenol-induced platelets inhibition were suspected, so the patient was immediately transferred to the Cardiac Surgery Department, where a gynecologists performed a caesarean section.

The patient gave birth to a boy (weight: 2110 g, Apgar score: 2/3/5/7 points).

After the procedure, the mother was in critical condition: cardiogenic and septic shock, tachycardia, severe acidosis. In addition, suprasystemic pressure in the pulmonary artery was noted. She required mechanical ventilation with 100% oxygen, intensive antibiotic therapy (meropenem, linezolid), as well as forcing diuresis. To maintain cardiovascular function, it was necessary to use an infusion of inotropes (adrenaline, noradrenaline, milrinone) and PAH-specific wide-range therapy. Stabilization of the circulatory system was obtained, but without a noticeable improvement in the patient's condition. In addition, restrictions were encountered in the escalation of the epoprostenol infusion due to thrombocytopenia and the features of a hemorrhagic diathesis manifested by respiratory bleeding. Increasing respiratory failure and haemodynamic instability necessitated the emergency use of arteriovenous extracorporeal membrane oxygenation (ECMO). Mechanical support allowed to reduce the

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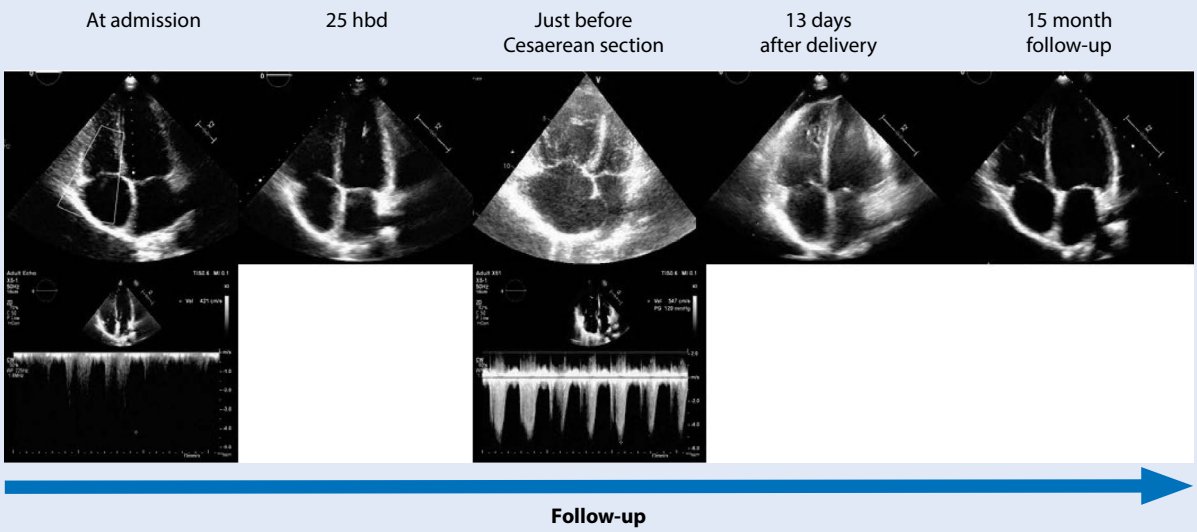

Figure 1. Transthoracic echocardiography — 4-Chamber views in different points of clinical follow-up

infusions of inotropes. Despite subsequent platelet substitution, the patient had another hemorrhagic complication, she required urgent laparotomy due to massive bleeding into the peritoneal cavity, and 3 days later relaparotomy was necessary because the bleeding returned. On the 7th day of its use, after adding levosimendan to therapy, it was decided to remove ECMO due to persistent bleeding. After 18 days post delivery, the patient returned to our ward, where pharmacotherapy consisted mainly of escalation of the infusion of epoprostenol. Ultimately, the escalation of therapy brought the expected clinical effect. In further treatment, a Hickman tunneled catheter was inserted into the left subclavian vein and a portable pump was used for intravenous epoprostenol administration.

At discharge it was recommended she use two methods of preventing pregnancy.

During the last hospitalization 15 months after delivery, the patient was in the WHO class I. She achieved treatment goals for PAH therapy on epoprostenol, macitentan, sildenafil and diltiazem treatment.

CONCLUSIONS

Current ESC guidelines [1, 2] advise against pregnancy in the presence of PAH as well as recommending pregnancy termination. Reactive form of PAH is related to less severe risk of pregnancy complications.

Initially, reactive iPAH was diagnosed in our patient. The patient responded well to diltiazem and sildenafil and the course of the first and second trimester of pregnancy was uncomplicated.

The dramatic course of the last pregnancy period was present. The final positive outcome resulted via the implementation of large amounts of inotropes, extracorporeal oxygenation and targeted treatment of PAH.

Regardless of the initial diagnosis of reactive iPAH, the patient necessitates the use of complex treatment to achieve the goals for PAH therapy.

In our opinion, the course of pregnancy in patients with reactive PAH may be unexpected, even dramatic, particularly in the third trimester of the pregnancy. In our opinion, it needs a coordinated multidisciplinary approach and may likely influence the long-term treatment and prognosis of the patient.

Conflict of interest

None.

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