

# Successful right heart remodelling and subsequent pregnancy in a patient with chronic thromboembolic pulmonary hypertension undergoing balloon pulmonary angioplasty: a case report

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## Background

To date, the management of pregnancy in patients with chronic thromboembolic pulmonary hypertension (CTEPH) and the associated risk of maternal mortality have not been established. Although balloon pulmonary angioplasty (BPA) in patients with CTEPH is still an emerging procedure, this approach represents a promising alternative to pulmonary endarterectomy (PEA), especially in patients with inoperable CTEPH.

## Case summary

We present a case of a 34-year-old woman with CTEPH who desired to have a child. Right heart catheterization showed a mean pulmonary artery pressure of 54 mmHg. Since the lesions were observed in the distal part of sub-segmental pulmonary arteries, there was no indication for PEA. After improvement in her haemodynamic status by BPA, she became pregnant. At 40 weeks of gestation, a normal baby was delivered vaginally. Both mother and baby have made satisfactory progress.

## Discussion

In cases in which the haemodynamic status is improved by effective BPA, pregnancy and childbirth may be possible, even in patients with CTEPH.

## Keywords

Chronic thromboembolic pulmonary hypertension (CTEPH) • Balloon pulmonary angioplasty (BPA) • Pregnancy • Vaginal delivery • Case report

## Learning points

- Balloon pulmonary angioplasty (BPA) is a promising alternative to pulmonary endarterectomy for distal-type chronic thromboembolic pulmonary hypertension (CTEPH).
- In cases where haemodynamic status is improved by effective BPA, pregnancy and childbirth may be possible, even in patients with CTEPH.
- Strict echocardiography follow-up should be performed during pregnancy.

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## Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) is defined as Group 4 pulmonary hypertension (PH) caused by organized thrombi. The prognosis is miserable, if CTEPH is untreated.<sup>1,2</sup> Pulmonary endarterectomy (PEA) is an established and curative surgical intervention for the proximal type of CTEPH.<sup>3</sup> However, the surgical approach for the distal type of CTEPH is difficult,<sup>4</sup> and a previous paper reported that only 56.8% of patients with CTEPH underwent PEA.<sup>5</sup> Although balloon pulmonary angioplasty (BPA) in patients with CTEPH is still an emerging procedure, this approach represents a promising alternative to PEA, especially in patients with inoperable CTEPH.<sup>6–8</sup> Pregnancy in women with pulmonary arterial hypertension is known to be associated with a high maternal mortality that is estimated to occur in 30–56% of cases.<sup>9</sup> To date, the management of pregnancy in patients with CTEPH and the associated risk of maternal mortality have not been established.

## Timeline

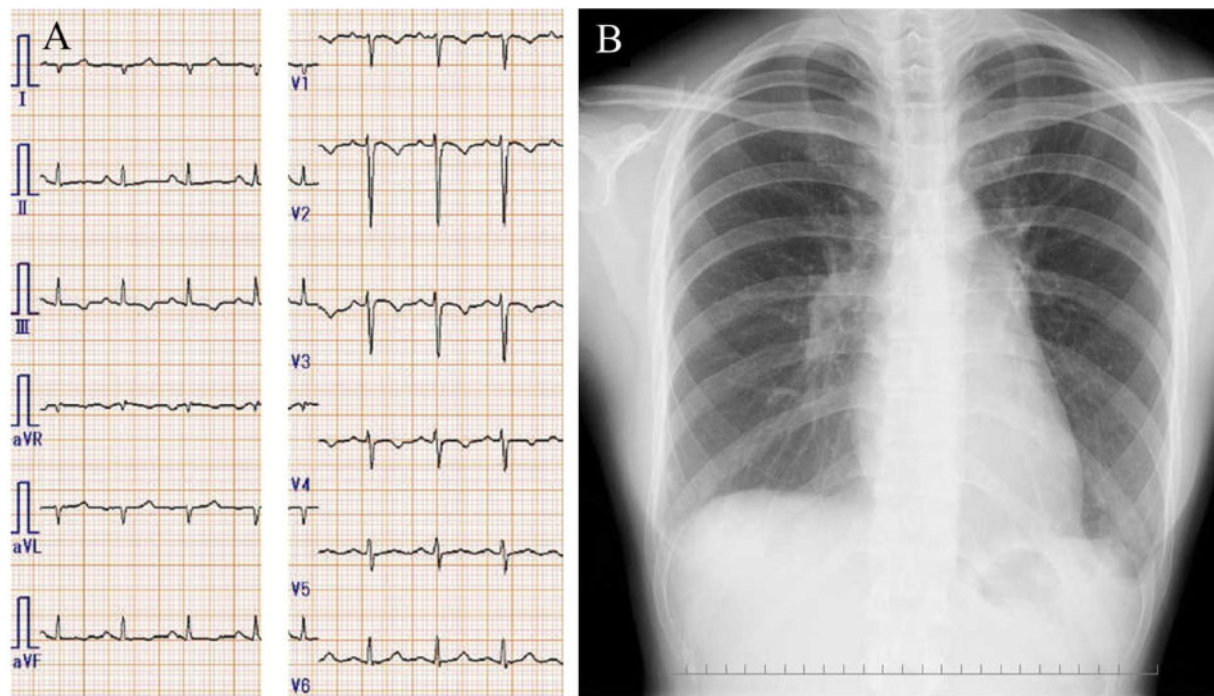
Time	Events
Initial presentation (Day 1)—1 week	A 34-year-old woman. Presented with a 4-month history of shortness of breath. Suspected chronic thromboembolic pulmonary hypertension (CTEPH) based on symptoms, history, blood work, chest X-rays, electrocardiogram, transthoracic echocardiogram, computed tomography, and perfusion scintigraphy.
2–3 weeks	Oral anticoagulant therapy (Rivaroxaban 15 mg/day). Right heart catheterization (RHC) revealed severe pulmonary hypertension and CTEPH specific angiographic images. The mean pulmonary artery pressure (mPAP) was 54 mmHg. The patient expressed a strong desire to have children.
1 month	No improvement of right heart load after 1 month of oral anticoagulant therapy. Ventricular tachycardia was observed, and emergency balloon pulmonary angioplasty was performed.
2 months	Balloon pulmonary angioplasty (six sessions) improved mPAP to 18 mmHg.
6 months	Oral anticoagulant therapy (rivaroxaban 15 mg/day). Follow-up RHC confirmed improvement of mPAP (19 mmHg) was maintained.
9 months	Heparin calcium subcutaneous injection (target activated partial thromboplastin time was 50–65 s). RHC mPAP 21 mmHg.
11 months	Heparin calcium subcutaneous injection 8750 U b.i.d. Pregnancy was confirmed.
1 year and 6 months	Successful vaginal delivery at 40 weeks of gestation.

## Case presentation

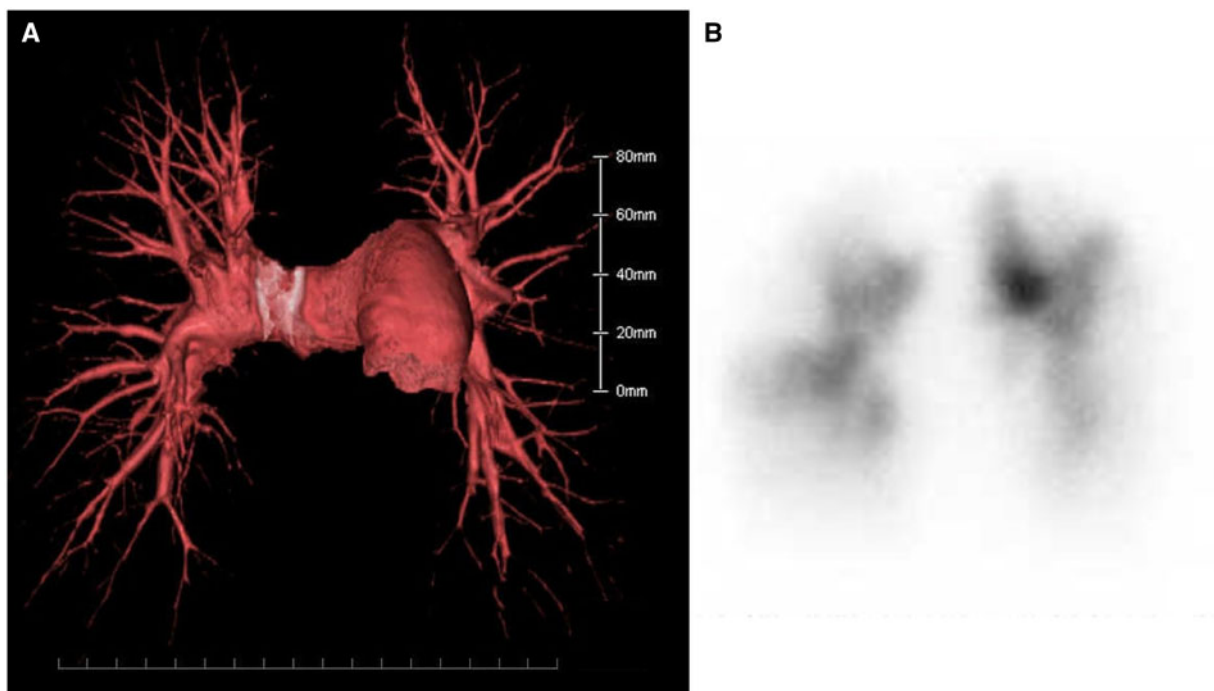
A 34-year-old woman was referred to our institution with a 4-month history of shortness of breath during exertion. She did not have a significant past medical history and no previous pregnancy or childbirth. The blood pressure was 106/72 mmHg, the heart rate was regular at 107 b.p.m., and the respiratory rate was 18/min. Although the oxygen saturation (on room air) was 95%, it decreased to 80% with low-level activity. An electrocardiogram showed right axis deviation and an inverted T in leads V1–3 (Figure 1A). A chest X-ray revealed dilation of the pulmonary artery (Figure 1B). A transthoracic echocardiogram revealed severe PH with a tricuspid systolic pressure gradient of 87 mmHg, an estimated mean pulmonary artery pressure (mPAP) of 49 mmHg, significant right atrial and right ventricular enlargement and left ventricular oppression by the right ventricle. Computed tomography presented a mosaic perfusion pattern and multiple chronic or organized thromboembolic obstructions in the distal part of the pulmonary arteries (Figure 2A). Perfusion scintigraphy showed severe perfusion defects (Figure 2B). Right heart catheterization (RHC) showed a mPAP of 54 mmHg (normal reference value: mPAP ≤ 20 mmHg), and pulmonary angiography revealed angiographical findings consistent with the distal type CTEPH. There was no finding suggesting an underlying coagulopathy in her blood tests. Although the patient had a strong wish to have a child, we prioritized treatment of CTEPH and started anticoagulant therapy (Rivaroxaban 15 mg/day).

Despite the continuation of anticoagulant therapy, there was no improvement in the right heart load as estimated by echocardiography and by her shortness of breath. On the 28th day, ventricular tachycardia caused by right heart overload was observed, and an emergency BPA was performed. After a total of six BPA sessions, the mPAP improved to 18 mmHg (Figure 3). Since the lesions were observed in the distal part of subsegmental pulmonary arteries, there was no indication for PEA.

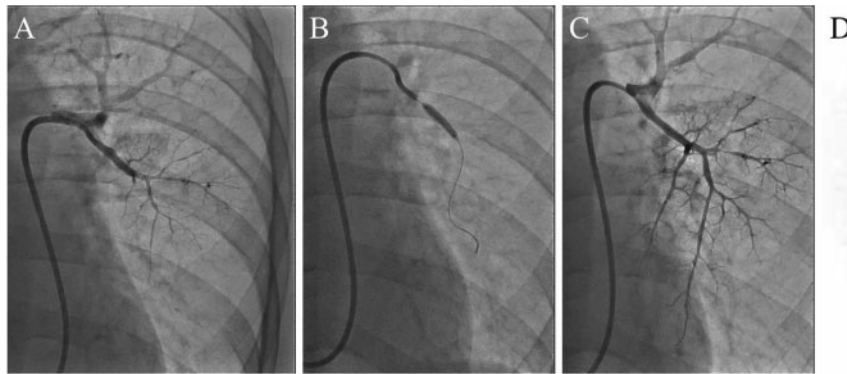
After continuing anticoagulation therapy for 3 months, RHC confirmed that there was no deterioration in the PH. Even at this point, the patient's strong wish to have a child had not changed, and she accepted the risk to her life that would result from pregnancy. Therefore, we changed the anticoagulation treatment from rivaroxaban to heparin calcium (subcutaneous injection). We maintained the activated partial thromboplastin time (APTT) between 50 and 65 s for another 3 months. After 3 months of heparin treatment, RHC was performed again, and it was confirmed that her haemodynamic condition had not deteriorated (mPAP 21 mmHg). Two months after the last RHC, pregnancy was confirmed. Heparin calcium subcutaneous injection was continued during the pregnancy, and the APTT was maintained between 50 and 65 s. No exacerbation of right heart load was observed by echocardiography at the follow-up examinations that were conducted every 2 months (Figure 4). At 40 weeks of gestation, a normal baby (3450 g) was delivered vaginally following the cessation of heparin calcium the preceding day and during the day of delivery, without any complications. Echocardiography revealed no exacerbation of PH after delivery. Both mother and baby have made satisfactory progress. She was enjoying motherhood (at the most recent follow-up, which was 10 months following delivery).



**Figure 1** A 12-lead resting electrocardiogram (A) and chest X-ray (B) on admission.



**Figure 2** Three-dimensional reconstructed computed tomography image showing multiple chronic or organized thromboembolic obstructions in the distal part of the pulmonary arteries (A). Perfusion scintigraphy revealed severe perfusion defects (B).



**Figure 3** Balloon pulmonary angioplasty (A: before treatment, B: balloon dilatation, C: after treatment) and perfusion scintigraphy after six sessions of balloon pulmonary angioplasty (D).

## Discussion

Although the treatment and management of PH have improved in recent decades, the maternal mortality rate remains high. In 2018, European Society of Cardiology (ESC) Guidelines for the management of cardiovascular diseases during pregnancy stated that pregnancy is not recommended (Class III, Level B) for patients with pulmonary artery hypertension (Group 1 PH). Furthermore, contraception should be advised, and termination should be considered if pregnancy occurs.<sup>9–13</sup>

Although PEA is a curative treatment for proximal CTEPH types, there are no established data regarding pregnancy and delivery of patients who underwent PEA because pregnancy causes hypercoagulability and increases the risk of thrombosis.<sup>1–3,14</sup> Furthermore, there has been no report concerning pregnancy and delivery after BPA. There is no standard thromboprophylaxis strategy for pregnant women with PH, and low-dose subcutaneous heparin is generally recommended.

In pregnant women with pulmonary artery hypertension, impaired prostacyclin production decreases the tolerance to large volume loads in the pulmonary circulation and increases pulmonary artery pressure.<sup>15</sup> Whether this phenomenon also applies to CTEPH is unknown. Therefore, we strictly followed the patient with serial echocardiography during pregnancy.

In this case, vaginal delivery was chosen. The optimal mode of delivery for PH patients remains controversial. Recently, the rates of caesarean sections have increased.<sup>12</sup> However, compared with caesarean sections, vaginal delivery is associated with less bleeding, fewer clotting complications, smaller blood volume shifts, and lower risks of infection. Because CTEPH is a thromboembolic disease, vaginal deliveries that have a lower risk for venous thromboembolism can be an attractive option, especially for pregnant women whose clinical condition of CTEPH is stable, as in the present case. Caesarean sections would be limited to situations in which there is a concern for a deterioration in maternal haemodynamics with the stress of active labour in cases of poorly controlled CTEPH.

This case is the first report of a patient with CTEPH who became pregnant and gave birth after improvement in her haemodynamic

status by BPA. In cases in which the haemodynamic status is improved by effective BPA, pregnancy and childbirth may be possible, even in patients with CTEPH.

## Lead author biography



Nobutaka Ikeda, MD, PhD, was born on 27 January 1977. He is from the Division of Cardiovascular Medicine at Toho University Ohashi Medical Center in Tokyo, Japan. He received his MD from the Shinshu University, Japan in 2001 and PhD from the Toho University, Japan.

## Supplementary material

**Supplementary material** is available at *European Heart Journal - Case Reports* online.

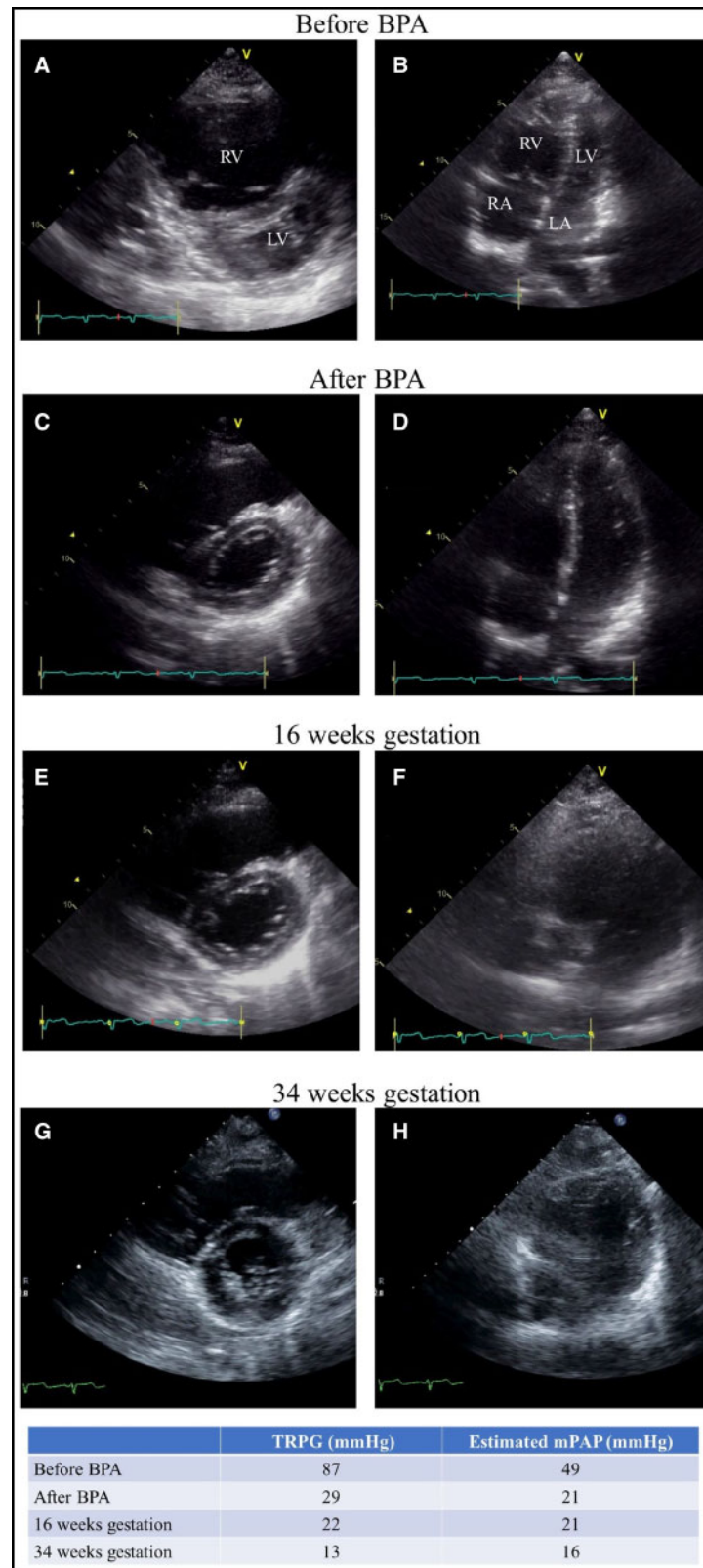
**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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**Figure 4** Short-axis view and apical four-chamber view of transthoracic echocardiogram before balloon pulmonary angioplasty (A, B), after balloon pulmonary angioplasty (C, D), at 16 weeks of gestation (E, F) and at 34 weeks of gestation (G, H). Apical four-chamber view images (F, H) were poor during pregnancy.

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