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BEGINNER

CASE REPORT: CLINICAL CASE

Pulmonary Embolism After Vaginal Delivery in a Fontan Patient

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ABSTRACT

The Fontan procedure was created to address the mixing of pulmonary and systemic venous return in patients with a single functional ventricle. The patient in this case with a Fontan repair experienced multiple pulmonary emboli 10 days post-partum. We outline management and recommendations when treating these patients. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:1713–5) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

PRESENTATION

This patient was a 20-year-old single pregnancy (G1) with viable offspring (P1) and 1 abortus (001) status post vacuum-assisted vaginal delivery at 39 weeks. During her pregnancy, she was receiving amoxicillin prophylaxis for asplenia prophylaxis, 250 mg daily, and aspirin, 81 mg daily. Her peripartum period was complicated by presumed chorioamnionitis/endometritis and was treated with ampicillin, gentamicin, and clindamycin. Additionally, she had a post-partum hemorrhage with estimated blood loss of 3 liters and underwent a dilation and curettage due to concern for retained products. She was discharged 4 days post-partum with plans for follow-up in 2 weeks. She presented 10 days post-partum with shortness of breath and right-sided chest tightness which worsened with inspiration since the previous day. Upon arrival she was hypoxemic with o_2 saturation of 50% to 60% and tachycardia above 150 beats/min. Physical examination revealed labored breathing and decreased breath sounds in the lower lung lobes bilaterally.

LEARNING OBJECTIVES

- To recognize increased risk for VTE in Fontan patients in pregnancy.
- To discuss current guidelines for VTE prevention in Fontan patients and pregnancy.

MEDICAL HISTORY

This patient had a history of heterotaxy syndrome (right isomerism type) with a double-inlet left ventricle, multiple ventricular septal defects, pulmonary atresia, and bilateral superior vena cava status post Fontan procedure. Her surgical history included a Blalock-Taussig shunt, bilateral Glenn anastomosis, and lateral tunnel fenestrated Fontan, followed later by a percutaneous closure of the Fontan fenestration. Echocardiography showing normal

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ABBREVIATIONS AND ACRONYMS

VE = venous thromboembolism

systolic function and mild atrioventricular valve regurgitation was noted in the first and third trimesters.

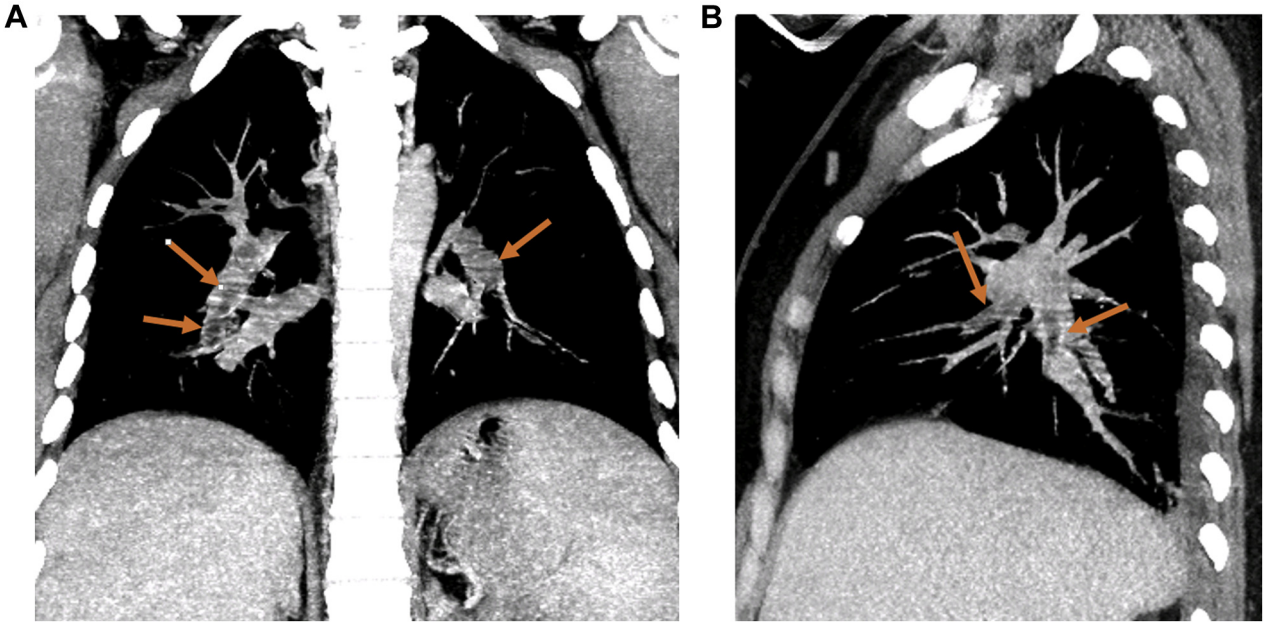
DIFFERENTIAL DIAGNOSIS AND INVESTIGATIONS. Given the patient's medical history of a Fontan procedure and her presentation of acute, pleuritic chest pain with low oxygen saturation upon admission, it was suspected that the patient experienced a pulmonary embolism (PE). Peripartum cardiomyopathy and acute myocardial infarction were also considered as possible diagnoses. Upon arrival, a complete blood count, complete metabolic panel, brain natriuretic peptide, and troponin level tests were ordered. Troponin concentration was normal; brain natriuretic peptide was elevated (133.1 pg/ml), but peripartum cardiomyopathy was ruled out because chest radiography results were negative for cardiomediastinal changes, infiltrates, and pleural effusion. Her chest computed tomography scan result was positive for nearly occlusive emboli within the right lower lobar artery and a nearly occlusive thrombus in the right upper lobar artery (Figure 1). Multiple smaller thrombi were noted. Transthoracic echocardiography demonstrated no pericardial effusion and no abnormal left ventricular function. Lower extremity Doppler readings were negative.

MANAGEMENT. Therapeutic enoxaparin was administered prior to her computed tomography scan due to the presumed diagnosis of PE. On the second day of admission, she underwent left heart catheterization for evaluation of Fontan hemodynamics and cardiac output in the context of large clot burden. She also underwent right heart catheterization and placement of Ekos catheters (ColoPlast, Fredensborg, Denmark) bilaterally in the right and left main pulmonary arteries with alteplase (tissue plasminogen activator) for 12 h. The Ekos catheters were removed on the second post-operative day, and daily warfarin, 5 mg, was started. She was discharged home on hospital day 5 on 2 l of oxygen with 6 months of anticoagulation therapy on warfarin with bridging with enoxaparin with the INR value goals of 2 to 3.

DISCUSSION

The Fontan procedure addresses the mixing of pulmonary and systemic venous return in patients with a single functional ventricle (1). The procedure redirects blood returning from the systemic circulation from the superior and inferior vena cavae to the pulmonary arteries, thereby bypassing the right ventricle (2). Pregnancy increases risk of

FIGURE 1 Maximum Intensity Projection Computed Tomography Images



Coronal **(A)** and sagittal **(B)** maximum intensity projection computed tomography images demonstrate filling defects in bilateral lower lobar and segmental and right middle lobe segmental pulmonary arteries, consistent with acute thromboemboli. The examination was performed in the equilibrium phase, given the history of prior Fontan palliation.

thromboembolic events as it induces a hypercoagulable state (3,4). These changes may not normalize until 8 weeks after delivery and lead to an increased risk of arterial and venous thromboembolism (VTE) during pregnancy and post-partum (3).

Previous studies indicate a prevalence for thromboembolic events in Fontan patients of 1% to 30% (5). However, guidelines for anticoagulation in Fontan patients are evolving (5). One study showed no significant differences in thromboembolic events for Fontan patients treated with antiplatelet therapy compared to well-regulated anticoagulation therapy (5). American Heart Association guidelines recommend antiplatelet therapy after the Fontan procedure but anticoagulation only in high-risk patients (6). European Society of Cardiology guidelines indicate that Fontan pregnancies are moderate to high risk for thromboembolism and that anticoagulation should be considered (7).

In a study by Pundi et al. (8), 70 pregnancies were reported in Fontan patients. Although patients in that cohort experienced other complications, none experienced thromboembolic events during pregnancy, despite the fact that only 63% of patients in that cohort were taking anticoagulation therapy, and 16% were taking antiplatelet therapy (8). The present study also used Ekos catheters to treat PE in the present patient. A study by McCabe et al. (9) demonstrated the effectiveness of Ekos catheters for treatment of PE. In that study, 53 patients with PE were treated using ultrasonography-assisted catheter-directed thrombolysis. Patients experienced reductions in pulmonary artery pressures after the procedure (9).

FOLLOW-UP

Within a month of follow-up, the present patient discontinued use of home oxygen as she had reached her baseline in the low 90s. Her warfarin dosage was adjusted to 6.5 mg daily to reach her INR goal of 2 to 3. Six months into warfarin therapy, she stopped breastfeeding and was transitioned to apixaban, 5 mg twice daily. Although evidence for the use of apixaban for recurrent PE prophylaxis is limited, a study by Georgekutty et al. (10) indicated that apixaban can be effective in Fontan patients with a history of thrombosis.

CONCLUSIONS

Although the present patient was treated during pregnancy with low-dose aspirin in accordance with American Heart Association guidelines, she experienced post-partum pulmonary emboli. Current guidelines from the European Society of Cardiology suggest anticoagulation should be considered in Fontan patients during pregnancy if there are other risk factors for VTE. Similarly, American Heart Association guidelines recommend that anticoagulation should be considered in Fontan patients who are at-risk (see discussion above). The venous stasis that occurs in the single ventricle along with the increased thrombotic risk during the pregnancy, specifically in the post-partum period, suggests that the Fontan may itself be a significant risk factor for VTE. The present case report adds to the studies of risk for VTE in patients with Fontan and pregnancy. In patients without contraindications, prophylactic anticoagulation may be considered for Fontan patients at 36 weeks' pregnancy and for 12 weeks' post-partum.

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KEY WORDS congenital heart defect, pregnancy, thrombus