中文題目：產後急性大範圍肺栓塞以非低血氧性呼吸衰竭表現--一蛋白S 缺乏之個案報告

英文題目：Postpartum acute massive pulmonary embolism presenting with nonhypoxemic respiratory failure: a case report with protein S deficiency

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Background: Protein S (PS), a vitamin K-dependent glycoprotein, is a cofactor of the protein C system. In the presence of PS, activated protein C inactivates factor Va and factor VIIIa, resulting in reduced thrombin generation. PS also serves as a cofactor for protein C enhancement of fibrinolysis, and can directly inhibit prothrombin activation. During pregnancy and in the postpartum period, functional protein S levels are significantly reduced when compared to nonpregnant females. In an analysis of 71 protein S-deficient members from 12 Dutch pedigrees, 38 percent of the individuals sustained pulmonary embolism. Herein, we reported a protein S-deficient postpartum woman with acute massive pulmonary embolism initially presenting with nonhypoxemic respiratory failure.

Case report: This 37-year-old G2P1woman (P1 via NSD, full term) presented with acute onset of fainting and shock two days after delivering a baby by cesarean section. The past history was unremarkable except gestational diabetes mellitus. Endotracheal intubation, fluid resuscitation, mechanical ventilation, and inotropic agents were prescribed. The arterial blood gas revealed 440 mmHg on PaO2 while 100 % oxygen was supplied by mechanical ventilator. Bedside echocardiography revealed right ventricle dilatation, severe tricuspid regurgitation, pulmonary hypertension (RVSP=55 mmHg), and acute left ventricle global hypokinesia. Therefore, ECMO was prescribed for cardiogenic shock. Electrocardiogram showed S1Q3T3 pattern in limb leads. The chest radiograph demonstrated left pulmonary artery engorgement with Knuckle sign present. Acute bilateral massive pulmonary embolism was confirmed by filling defect within pulmonary trunk, bilateral pulmonary artery, left superior and inferior pulmonary artery, and right inferior pulmonary artery. Emergent thrombus aspiration, instead of thrombolytic therapy owing to post-cesarean section, was performed successfully by pulmonary angiojet catheter. Anticoagulant therapy was prescribed for the residual thrombus, but transarterial embolization was performed due to complicated uterine artery bleeding. Finally, she was weaned from ECMO, mechanical ventilator, and inotropic agents. Oral form anticoagulant therapy was kept on after delivery and a thorough survey for pulmonary embolism was employed. Protein S deficiency (protein S:13.9%; reference rage 60-130%) was impressed.

Discussion: Acute pulmonary embolism (PE), often high mortality rate of approximately 30 percent without treatment, is very difficult to diagnosis and manage on postpartum women with reduced functional protein S levels. In our case, first, the typical arterial blood gas findings are not always seen. As an example, massive PE with hypotension and respiratory collapse can cause hypercapnia and a combined respiratory and metabolic acidosis. In addition, hypoxemia can be

minimal or absent. A PaO2 between 85 and 105 mmHg exists in approximately 18 percent of patients with PE. Up to 6 percent may have a normal alveolar-arterial gradient for oxygen. Second, during pregnancy and in the postpartum period, functional protein S levels are significant reduced (38% +/- 17.3%, mean +/- 1 SD) when compared to nonpregnant females (97% +/- 31.6%) ($P < 0.001$) due to total protein S levels are reduced (68% +/- 10.7%) compared to nonpregnant controls (100% +/- 17.0%) ($P < 0.001$). Although several case reports have described young patients with arterial thrombosis and hereditary protein S deficiency, however, larger studies have not convincingly demonstrated that protein S deficiency is a risk factor for the development of arterial thrombosis. Third, no reports regarding to the treatment of acute pulmonary embolism in postpartum women with functional protein S deficiency were published in the literature review. Therefore, she is temporarily treated with oral anticoagulant drugs.

Conclusion: Acute massive pulmonary embolism on postpartum women with functional protein S deficiency is difficult to diagnosis and often lethal. We should take it into consideration even it presents with nonhypoxemic respiratory failure. As to the treatment, it warrants further studies.