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## CLINICAL INFORMATION

# Anesthesia for cesarean section in a patient with isolated unilateral absence of a pulmonary artery



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### KEYWORDS

Anesthesia;  
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### Abstract

**Background and objectives:** Congenital unilateral absence of a pulmonary artery (UAPA) is a rare anomaly. Although there are several reports regarding pregnancy in patients with unilateral absence of a pulmonary artery, there are no case reports describing anesthesia for Cesarean section in a patient with unilateral absence of a pulmonary artery.

**Case report:** We present a patient with unilateral absence of a pulmonary artery who underwent Cesarean sections twice at the ages of 24 and 26 years under spinal anesthesia for surgery and epidural analgesia for postoperative pain relief. Both times, spinal anesthesia and epidural analgesia enabled successful anesthesia management without the development of either pulmonary hypertension or right heart failure.

**Conclusion:** Spinal anesthesia combined with epidural analgesia is a useful anesthetic method for a Cesarean section in patients with unilateral absence of a pulmonary artery.

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### PALAVRAS-CHAVE

Anestesia;  
Cesariana;  
Gravidez;  
Ausência unilateral  
de uma artéria  
pulmonar

### Anestesia para cesariana em paciente com ausência unilateral isolada de artéria pulmonar

### Resumo

**Justificativa e objetivos:** A ausência congênita unilateral de uma artéria pulmonar (ACAP) é uma anomalia rara. Embora existam vários relatos sobre pacientes grávidas com ACAP, não há relatos de casos descrevendo anestesia para cesariana em pacientes com ACAP.

**Relato de caso:** Apresentamos uma paciente com ACAP que foi submetida a duas cesarianas, nas idades de 24 e 26 anos, sob raqui-anestesia para a cirurgia e analgesia epidural para a dor no pós-operatório. Nas duas cesarianas, a raqui-anestesia e analgesia epidural possibilitaram

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o manejo bem-sucedido da anestesia, sem a ocorrência de qualquer hipertensão pulmonar ou insuficiência cardíaca direita.

**Conclusão:** Raquianestesia combinada com analgesia epidural é um método anestésico útil para cesarianas em pacientes com ACAP.

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## Introduction

Congenital unilateral absence of a pulmonary artery (UAPA) is a rare anomaly with an estimated prevalence of approximately 1 in 200,000 young adults.<sup>1</sup> Patients who have no cardiac anomalies other than UAPA can remain asymptomatic even into late adulthood. The most common symptoms are recurrent pulmonary infections, decreased exercise tolerance or mild dyspnea on exertion.<sup>2</sup> The symptoms of isolated UAPA can be provoked by predisposing factors, such as pregnancy<sup>3-8</sup> or high altitude.<sup>2</sup> Pregnancy is known to increase cardiac output. Furthermore, unilateral lung perfusion with the entire cardiac output is a risk factor for the development of pulmonary arterial hypertension. The prognosis of isolated UAPA depends on the presence or absence of pulmonary arterial hypertension.<sup>2</sup> Although there are several reports regarding pregnancy in patients with UAPA,<sup>3-9</sup> there are no case reports describing anesthesia for a Cesarean section in a patient with UAPA. Thus, the best approach to anesthesia for Cesarean section in these patients remains unclear. We present a patient who underwent Cesarean sections under spinal anesthesia for surgery and epidural analgesia for postoperative pain relief twice at the ages of 24 and 26 years.

## Case report

### First cesarean section

A 24-year-old woman, pregnant patient, with UAPA was admitted to our hospital at 35 weeks' gestation for delivery and perinatal care. She was diagnosed with UAPA at the age of 15 years when she incurred right-sided pneumonia. A chest X-ray revealed the absence of the right pulmonary artery trunk. Computed tomography revealed the absence of the right pulmonary artery and the presence of three collateral vessels from the ascending aorta to the right lung. An echocardiogram did not image any additional cardiac anomalies. Right cardiac catheterization revealed normal pulmonary arterial pressure (PAP). At the time of diagnosis, she was asymptomatic for UAPA and surgical correction was not indicated. She had no past medical history other than UAPA and the right-sided pneumonia.

On admission, the patient was asymptomatic. She had no pregnancy-related complications. Her height was 154 cm and her weight was 63 kg. A chest X-ray showed the absence of the right pulmonary artery trunk, mediastinal shift to the right and an expanded left pulmonary artery trunk



**Figure 1** Chest roentgenography showing absence of the right pulmonary artery trunk, decreased pulmonary vasculature in the right lung, shift of the mediastinal structures to the right and an expanded left pulmonary artery trunk.

(Fig. 1). Chest magnetic resonance imaging revealed that blood flow in the left pulmonary artery was approximately twice the normal volume. Her PAP was normal. Arterial blood gas analysis revealed a pH of 7.48, oxygen pressure of 83.8 mmHg and carbon dioxide pressure of 34.7 mmHg when breathing air. Although she was initially scheduled to undergo a vaginal delivery under epidural analgesia with systemic and PAP monitoring, labor had not begun by 38 weeks and 4 days' gestation; therefore, a Cesarean section with stringent systemic and PAP monitoring was planned. A pulmonary artery flotation catheter was inserted on the day before surgery. On arrival to the operating room (OR), systemic arterial pressure (SAP) was 106/68 mmHg, PAP was 5/1 mmHg, heart rate (HR) was 95 beats/min and oxygen saturation (SpO<sub>2</sub>) was 98%. An arterial catheter was inserted in the OR. After administering oxygen at 5 L/min

via a face mask, an epidural catheter was inserted at the Th12/L1 interspace. Next, spinal anesthesia was administered at the L3/L4 interspace with 10 mg of hyperbaric bupivacaine hydrochloride hydrate. Five minutes after the injection, SAP was 108/52 mmHg, PAP was 6/2 mmHg, HR was 82 beats/min and SpO<sub>2</sub> was 99%. Fifteen minutes after the injection, the sensory block level was between Th1 and S5. At this time, 300 mL of crystalloid fluid and 400 mL of colloidal fluid had been administered intravenously. SAP, PAP and SpO<sub>2</sub> did not change significantly. No cardiovascular agents were administered. Intraoperatively as well, SAP, PAP and SpO<sub>2</sub> did not change significantly. At delivery, the neonate had Apgar scores of 8 and 9 at 1 and 5 min, respectively. During the delivery, the PAP increased slightly from 10/7 mmHg to 15/6 mmHg while the SAP increased slightly from 100/50 mmHg to 108/65 mmHg. After the placenta was delivered, 10 units of oxytocin were administered intravenously, which is routine practice at our institution. This administration caused a slight decrease in SAP from 108/65 mmHg to 100/55 mmHg and no changes in PAP. Midazolam was administered at a dose of 2 mg as a sedative, which resulted in no changes in SAP or PAP. The administration of a sedative was based on patient demand. Total blood loss excluding dilution with amniotic fluid was approximately 180 mL. Total fluids administered under PAP monitoring and central venous pressure (CVP) monitoring included 400 mL of crystalloid fluid and 900 mL of colloidal fluid. The patient was pain-free throughout the operation. No cardiovascular agents were administered. The surgery was completed uneventfully. Postoperative pain was adequately controlled by epidural analgesia with 0.2% ropivacaine at a rate of 5 mL/h, without supplementary analgesics. The highest pulmonary systolic pressure reached was 20 mmHg on the second postoperative day. Postoperatively, the clinical courses of the patient and her infant were uneventful.

## Second cesarean section

The patient achieved a second pregnancy at age of 26 years and was admitted to our hospital at 36 weeks' gestation with no pregnancy-related complications. Her height was 154 cm and her weight was 61 kg. Her PAP was within normal limits. She was scheduled to undergo Cesarean section at 37 weeks' gestation under the same anesthetic plan as that of her first Cesarean section. On arrival to the OR, SAP was 115/76 mmHg, PAP was 8/4 mmHg, HR was 90 beats/min and SpO<sub>2</sub> was 97%. After application of all monitors as before and administration of 5 L/min of oxygen via a face mask, an epidural catheter was inserted at the Th12/L1 interspace. Next, spinal anesthesia was administered at the L3/L4 interspace with 9 mg of hyperbaric bupivacaine hydrochloride hydrate. Five minutes after the injection, SAP was 115/52 mmHg, PAP was 10/5 mmHg, HR was 92 beats/min and SpO<sub>2</sub> was 100%. Fifteen minutes after the injection, the sensory block level was between Th2 and S5. At this time, 300 mL of crystalloid fluid and 300 mL of colloidal fluid were administered intravenously. SAP, PAP and SpO<sub>2</sub> did not change significantly. Therefore, no cardiovascular agents were administered. At delivery, the neonate had Apgar scores of 8 and 9 at 1 and 5 min, respectively. During the delivery, the patient's PAP increased slightly

from 11/4 mmHg to 15/7 mmHg. After the placenta was delivered, intravenous oxytocin at a dose of 10 units and intravenous midazolam at a dose of 2 mg did not cause any significant changes in SAP or PAP. Total blood loss excluding dilution with amniotic fluid was 190 mL. Total fluids administered under PAP monitoring and CVP monitoring included 800 mL of crystalloid fluid and 500 mL of colloidal fluid. No cardiovascular agents were administered. The patient was pain-free throughout the operation, which was performed uneventfully. Postoperative pain was adequately controlled with epidural analgesia with 0.2% ropivacaine at a rate of 5 mL/h, without supplementary analgesics. The highest pulmonary systolic pressure reached was 22 mmHg 2 h after the conclusion of surgery. The postoperative clinical courses of the patient and her infant were uneventful.

## Discussion

There have been no previous case reports describing anesthesia for Cesarean section in patients with UAPA. We administered spinal anesthesia for surgery and epidural analgesia for postoperative pain relief to our patient with UAPA. The advantage of this protocol is that it blocks intraoperative and postoperative nociceptive stimuli caused by surgery, thus preventing the cardiovascular changes induced by these nociceptive stimuli. The disadvantage is that patients might need vasopressor drug administration or a significantly large fluid infusion to maintain arterial pressure if the level of spinal block rises. During both surgeries in our patient, spinal anesthesia caused no significant cardiovascular or respiratory changes. Moreover, the patient did not develop any serious complications during the perioperative period.

Spinal anesthesia caused sensory blockade up to Th1 for the first surgery and Th2 for the second surgery. The possible reasons why the patient did not require any vasopressors included proper fluid administration and the patient's robust circulation. A slow incremental epidural anesthesia combined with the lower-dose spinal anesthesia was another anesthetic option for this case. This option could have regulated the region of sensory blockade.

The PAP increased slightly on the second postoperative day for the first surgery and 2 h postoperatively for the second surgery. It was probably due to ongoing uterine contractions after delivery. Uterine contractions induce hypervolemia especially during the first few postpartum days. Close observation of the PAP is important during this period. Therefore, postoperative analgesia is also important because nociceptive sympathetic stimulation might cause right heart overload.

Administration of 10 units of oxytocin did not cause any significant cardiovascular changes during both surgeries; however, this dose is large for an elective Cesarean section.<sup>10</sup> Adequate uterine contractions can usually be achieved with oxytocin at a dose of <5 units; this dosage can also minimize cardiovascular side effects.<sup>10-12</sup> Oxytocin is problematic particularly in patients with pulmonary hypertension because it raises the PAP.<sup>13</sup> In addition, the drop in systemic vascular resistance due to oxytocin.<sup>11</sup> can compromise the right ventricle. In this regard, in both surgeries, we should have discussed the dose of oxytocin with

obstetricians and used the minimum dose necessary for adequate uterine contractions while considering the amount of blood loss.

Although UAPA can remain asymptomatic until late adulthood, it can be unmasked by pregnancy,<sup>3-8</sup> probably due to increasing cardiac preload during pregnancy. Symptoms of UAPA can also be triggered by spinal anesthesia or Cesarean section, because these procedures might cause right heart failure or pulmonary hypertension. Right heart failure or pulmonary hypertension results from excessive cardiac preload or afterload due to increased venous return after delivery, excessive transfusion or excessive sympathetic stimulation due to nociceptive stimuli. Koga et al. reported a case of a woman in whom PAP rose after Cesarean section because of a sudden increase in venous return after labor.<sup>3</sup> In our patient, close cardiovascular and respiratory monitoring indicated that these factors were not significantly altered by either the spinal anesthesia or the Cesarean section. Spinal anesthesia for surgery and postoperative epidural analgesia resulted in complete pain relief. Thus, this protocol is a useful anesthetic option for Cesarean section in patients with UAPA.

A review by Harkel et al.,<sup>2</sup> reported a mortality rate of isolated UAPA to be 7 (6.5%) of 108. The cause of death included pulmonary hemorrhage, right heart failure, pulmonary hypertension, respiratory failure and high altitude pulmonary edema.<sup>2</sup> Pregnancy is known to increase cardiac output, which could induce pulmonary hypertension in pregnant patients with UAPA, leading to right heart failure or rupture of affected pulmonary capillaries. The literature contains only seven previous reports addressing the clinical course of pregnancy in patients with UAPA.<sup>3-9</sup> According to these studies, UAPA was unmasked by pregnancy in 10 patients who presented with symptoms such as dyspnea and chest pain.<sup>3-8</sup> One patient developed adult respiratory distress syndrome on the second postpartum day and died 15 days after delivery.<sup>6</sup> Therefore, patients with UAPA undergoing Cesarean section should be closely and continuously followed throughout the perioperative period, especially when uterine contractions are present.

## Conclusions

We administered spinal anesthesia for the Cesarean section and epidural analgesia for postoperative pain relief in

a patient with UAPA in two instances. In both instances, neither pulmonary hypertension nor right heart failure developed. Thus, this combination is a useful anesthetic method for Cesarean section in patients with UAPA.

## Conflicts of interest

The authors declare no conflicts of interest.

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