

# Anesthesia for Emergency Separation of Omphalopagus Conjoined Twins

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**Summary:** Hobaika ABS, Pires KCC, Fernandes VBSA – Anesthesia for Emergency Separation of Omphalopagus Twins.

**Background and objectives:** The rate of mortality during the surgical separation of conjoined twins in the neonatal period is 50% and can reach up to 75% if it occurs in emergency situations. The planning of the surgical separation procedure is detailed and involves imaging assessment, evaluation of cross-circulation and even other surgical preparation procedures, such as skin expansion.

**Case Report:** Eleven-day-old female omphalopagus conjoined twins underwent emergency surgical separation due to the death of one twin caused by sepsis associated to cardiopathy. The liver was shared by the twins and was separated. The surviving twin died six days later.

**Conclusions:** Surgical separation of conjoined twins in the neonatal period must be avoided due to the patients' organic system immaturity. However, emergency situations such as the one described herein can require the procedure to be carried out.

**Keywords:** ANESTHESIA, Pediatric; SURGERY, Pediatric: omphalopagus conjoined twins.

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## INTRODUCTION

It is estimated that conjoined twins occur in about one in every 100,000 pregnancies worldwide; however, most cases result in stillbirths or death shortly after birth. The separation surgery must be postponed until the infants are relatively mature (6 to 12 months) when survival rates reach 90%. The mortality rate in the neonatal period is 50%<sup>1-3</sup>. This case report describes the emergency surgical separation of omphalopagus twins due to the death of one of them.

## CASE REPORT

We report on the case of eleven-year-old female omphalopagus conjoined twins, born at term by vaginal delivery. Twin A presented cardiopathy (large patent ductus arteriosus, small ventricular septal defect and patent foramen ovale) and septic shock, which evolved to cardiorespiratory arrest and death after resuscitation maneuvers and intravenous adrenalin. Twin

B weighed 1.95 kg and presented early sepsis; at admission she presented tracheal intubation and catheter in the internal jugular vein and a heart rate of 198 bpm. Serum potassium was 4.3 and sodium was 137. The twins were sent for emergency surgical separation 40 minutes after the death of twin A. The team consisted of an anesthesiologist and a pediatric surgeon experienced in conjoined twin separation, two anesthesiologists that were part of the team and a resident in anesthesiology. Vecuronium (0,1 mg.kg<sup>-1</sup>) and midazolam (0.2 mg.kg<sup>-1</sup>) were administered and a caudal epidural anesthesia was performed with 100 µg of morphine and 4.0 mg of bupivacaine at L<sub>4</sub>/L<sub>5</sub>. Pulmonary ventilation of twin B was adjusted in order to keep P<sub>ET</sub>CO<sub>2</sub> at around 33 mmHg. Twin B was kept warm with a warming blanket. The separation surgery was uneventful and lasted 140 minutes during which a single and dysmorphic liver was identified. Heart rate was stabilized at 118 bpm, 45 minutes after the anesthetic induction. A total of 40 mL of red cell concentrate and 40 mL of plasma were administered. Twin B died on the 6th postoperative day due to septic shock.

## DISCUSSION

Omphalopagus conjoined twins comprise around 33% of the cases of conjoined twins and can vary from multiple-organ sharing to conjoined livers only <sup>1,4,6</sup>. The planning of the separation surgery through imaging assessment, evaluation of cross-circulation and even other surgical preparation procedures, such as skin expansion can be carried out.

In some situations the emergency separation surgery is indicated: intestinal obstruction, omphalocele rupture, con-

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gestive heart failure, obstructive uropathy, sepsis and difficult-to-treat respiratory or cardiovascular involvement<sup>2-6</sup>. In these circumstances, mortality is higher because the blood volume is lower (patients have not achieved the ideal age) and there is no time to plan the anesthetic-surgical procedure<sup>1-7,8</sup>.

Mabogoune et al. reported 12 cases of conjoined twins. Three cases underwent emergency surgical separation was performed, with a mortality rate of 75% and the causes of death were the sacrifice of a twin and cardiovascular collapse in two others<sup>8</sup>. Ure et al. reported the emergency separation of twins, of which one was apparently well-formed and the other was malformed who died during surgery. The surviving twin also died on the 7<sup>th</sup> postoperative day due to brain hemorrhage with coagulation disorder<sup>9</sup>. Spitz reports the experience of 17 cases of separation, of which 7 were emergency procedures (one due to the death of a twin) and 4 of the 14 twins survived (28%)<sup>10</sup>. Saguil et al. reported the emergency separation of 6 of 22 conjoined twins<sup>11</sup>. Of these 12 twins, only one survived. Graiveir et al. reported an emergency separation procedure due to the death of one of the twins due to respiratory failure<sup>7</sup>. Watanattitan et al. report a series of 11 cases of separation, of which 3 were emergency procedures; 1 due to the death of one of the twins – the other twin died after the separation (1 hour) and two others due to cardiovascular function deterioration in one of the twins (complex cardiac malformations)<sup>12</sup>. Jaffray B. et al. reported a case of omphalopagus in which one of the twins presented necrotizing enterocolitis treated through laparotomy, and the expectant conduct was chosen until the surgical separation could be performed<sup>13</sup>. However, the twin undergoing treatment deteriorated quickly, which led to the death of both twins. In this case the emergency separation was disregarded as it was considered a useless effort.

In the present case twin A presented multiple cardiac malformations, in addition to sepsis, which determined her clinical deterioration and death. One of the main causes of death in the postoperative period of the separation of conjoined twins

is cardiac malformation, as the adequate function of the cardiovascular system is necessary for a successful peri and postoperative period. In the case reported here the vascular bed of twin A worked as a volemic reserve, which remained vasoconstricted for some time due to effect of the adrenaline used during the resuscitation maneuvers. Therefore, twin B was very tachycardic. At the same time, the adrenalin that entered the circulation of twin B might have contributed to maintain her inotropic and chronotropic support preventing her immediate death. The cross-circulation seemed important and there was a concern that the hypoxic blood containing endotoxins and cytokines from twin A could put twin B at risk, triggering the activation of systems such as NOS-2, responsible for vasodilation, myocardial depression and alteration in the cellular calcium inflow, decreasing L-type calcium receptors<sup>14</sup>. These events might have contributed to the death of the surviving twin a few days later.

One can conclude that the emergency separation of conjoined twins is a high-mortality surgery (75%), determined by the causes that led to the separation, associated with the lack of time to plan the surgical-anesthetic procedure. At this age the organic systems are not yet mature: the heart is not prepared for hypervolemic or hypovolemic situations. The upper and lower airways have small-caliber, thorax compliance is high, the intercostal and diaphragmatic muscles have a higher content of fibers (type II) and lung compliance is low. The functional residual capacity is low and the volume of occlusion is high. That predisposes to higher respiratory work, onset of cyanosis and early fatigue of the respiratory muscles<sup>15-16</sup>. The kidney has difficulty retaining sodium under stress situations and there is tubuloglomerular imbalance, with more immature tubules. The liver has not developed the mechanisms to metabolize drugs. The concentration of proteins that bind to the drugs is small (albumin, acid  $\alpha < 1$ -glycoprotein), predisposing the patients to a higher percentage of free drugs<sup>15</sup>. The surgical separation of conjoined twins, therefore, must be carried out only in cases where there is risk of death<sup>7</sup>.