A set of 34-week omphalo-thoracopagus conjoined twins were prenatally diagnosed and followed at the Center for Fetal Diagnosis and Treatment-The Children’s Hospital of Philadelphia.

The diagnosis was made via fetal MRI and US. Other than a fused liver with complex vascular architecture and a shared pericardium Twin A had no other known anomalies. However, Twin B had severe cerebral ventriculomegaly, an absent corpus callosum, severe micrognathia, massive cardiomegaly and hypoplastic lungs. Of most importance, twin B had no aortic or pulmonary outflow—making him 100% dependent on Twin A for survival.

The operative plan was to deliver the twins by C-section, then proceed with immediate surgical separation. Due to the lack of pulmonary and aortic outflow in Twin B, there were no plans to manage him. The plan was to perform an awake intubation on Twin A and to establish intravenous and intra-arterial access.

The anesthesia teams consisted of one anesthesiologist who cared for the twins’ mother during the C-section. A second team consisted of two anesthesiologists and two fellows who cared for Twin A. Two anesthesia technicians were also available.

In preparation for massive blood loss, hemodynamic changes and ventilatory difficulty we had RBC’s, FFP and platelets in the operating room. Resuscitation drugs, including Factor VII, isoproterenol and inhaled nitric oxide were also readily available.

After delivery, the twins were brought into an adjacent operating room. Intubation proved challenging given the copious amounts of amniotic fluid in Twin A’s mouth. After appropriate intravenous access was secured, Twin B’s body was steriley prepped into the surgical field while Twin A was steriley prepped to the axilla.

Surgical separation lasted 3.5 hours. Findings confirmed preoperative imaging: a common pericardial sac, a connection between the small intestines, a shared liver, and anomalous cardiac architecture without cardiac outflow tracts in twin B.

Twin B’s entire arterial perfusion originated from an artery that came off Twin A’s aorta. This artery transversed the liver and went into Twin B’s hypoplastic aorta. There was one umbilical vein that entered the liver and then separated into two—one towards Twin A and the other into Twin B.

When the vessels between the twins were clamped, immediate cessation of blood flow resulted in cardiac standstill in Twin B. Twin A was expeditiously separated from Twin A. EBL was 100 mL. At the end of the procedure there were no electrolyte abnormalities. The final ABG was: pH 7.448, pCO2 35.6, pO2 43, BE +1, Hg 11.2.

Upon arrival to the NICU ABG revealed mixed metabolic and respiratory abnormalities that were treated. Over the next 48 hours the respiratory and metabolic abnormalities resolved. Twin A was extubated on postoperative day 15 and is overall doing well. The successful implementation and outcome of this case depended on careful multidisciplinary preoperative planning and coordination.