Early Separation of Thoraco-Omphalopagus Conjoined Twins: An Exercise in Preoperative Planning and Preparation

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Introduction
Conjoined twins occur once every 200,000 live births following uniovular gestation and incomplete fission of the embryonic disk at the 15-17 day stage. They are classified according to the most prominent site of union followed by the Greek suffix ‘pagus,’ meaning fixed. The most common types are thoracopagus (40%), omphalopagus (32%), pygopagus (19%), ischiopagus (6%) and craniopagus (2%). Management falls into three broad categories:
1. Non-operative management: Inoperable due to extensive or complex union, or inseparable hearts.
2. Elective separation: This can be early, as in the first few days of life, or delayed until the twins are several months or even years old.
3. Emergency separation: This may be necessary as a result of cardiopulmonary decompensation, bowel obstruction, or sepsis affecting one or both twins.

Case Report: We describe the separation of a set of female thoraco-omphalopagus conjoined twins on Day 5 of life. We discuss the anesthetic management of the twins for diagnostic imaging and separation in relation to their anomalies that included a shared hepatobiliary system, a fused duodenum, and fused chest and abdominal walls. We highlight the challenges associated with forced early separation due to bowel obstruction, and the postoperative management.

Discussion: Meticulous planning with 2 separate anesthesia teams and separate color-coded equipment for each twin is necessary for all stages of management. Cardiopulmonary decompensation may occur at any time, particularly with thoracopagus twins, hence the need for early planning in case of urgent separation. Airway difficulty is more common in twins with high thoracopagus and a close face-to-face position. Early separation in the neonatal period was indicated in these twins because of bowel obstruction. Operative survival is 50% in the neonatal period compared to 90% when >4m of age, and optimal separation time is between 4 -11 months. Elective delayed separation allows placement of tissue expanders to achieve primary skin closure of defect, reduces anesthetic risk, and allows confirmation or detection of anomalies.

Prenatal EXIT procedures can be useful to accurately evaluate the anatomic connections and congenital anomalies by echocardiography. EXIT may predict potentially fatal cardiac deterioration at birth that can then be managed in a controlled fashion. Associated anomalies are common even in organs that are not conjoined, and may affect survival even in twins deemed ‘separable’.