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Anesthetic Management of Thoracopagus Twins Undergoing Cardiopagus Separation

DAVID L. BROWN, M.D.,* DEBORAH M. HOLUBEC, M.D.,* DANIEL J. TOWLE, M.D.,†
A. KIRK BODARY, M.D.,† ALAN R. PATTERSON, M.D.,* JOHN MACK, M.D.‡

The first successful separation of conjoined twins, xiphopagus type (twins joined in the region of the xiphoid process), was carried out by Konig in 1690.[1] Xiphopagus twins born in Siam (Thailand) in 1811 have provided us with the common parlance of "siamese twins."[2] Several reviews have outlined the anesthetic management of separation procedures for conjoined twins,[3–5] one of which dealt exclusively with thoracopagus twins.[6]

Thoracopagus twins (twins joined in the thoracic region) represent 75% of conjoined twins.[7,8] In the past, attempts at separation were rare due to the high incidence (75%) of associated cardiac abnormalities.[9] From 1925 until 1970, only 10 attempts at separation of thoracopagus twins were recorded.[10] In 1977, Patel *et al.*[9] reported four attempts at thoracopagus separation with no survivors. Separation procedures involving thoracopagus–cardiopagus twins (twins sharing common myocardium) are even more rare. The single survivor of cardiopagus separation was a twin who shared only an interatrial vascular communication and had normal cardiac anatomy when the repair was completed.[11] In 1980 another attempt at thoracopagus, cardiopagus twin separation, also involving an interatrial vascular communication, was reported.[12] Neither infant from this procedure lived beyond 48 h postoperatively.

We describe the anesthetic and cardiovascular management of the first known attempt at separation of thoracopagus–cardiopagus twins, whose cardiac lesion involved both atria and ventricles.

Report of a Case

Preoperative Evaluation. Female conjoined twins at 35 weeks' gestation were born to a 34-year-old gravida 4, para 2 mother. The conjoined relationship of the 4.1-kg twins was documented by ultrasonography prior to their elective cesarean section birth. Both infants were cyanotic immediately after delivery, and a severe cardiac defect was suspected when tracheal intubation and ventilation of one infant resulted in improved color of both. The trachea of the second twin subsequently was intubated and ventilation managed in the neonatal intensive care unit (NICU).

Cardiac echocardiography and catheterization were performed the following day. This evaluation revealed a shared, common four-chambered heart and conduction system (fig. 1). A pulmonary artery and aorta arose from each ventricle. The infant on the right (baby B) had d-transposition of the aorta and pulmonary artery. The infant on the left (baby A) had severe pulmonary stenosis but anatomically correct great vessels. Atrial and ventricular septal defects were present. Both superior vena cavae drained to the appropriate atria. Inferior vena caval flow from both babies drained to the (baby A) left atrium. Pulmonary venous return was indeterminant at catheterization, but at operation, baby B's pulmonary return was to the topographic right atrium (baby B) and that of baby A to the topographic left atrium (baby A) (fig. 1). The infants preoperatively were suspected of having a common liver, while the remaining organ systems were thought to be separate.

Conjoined survival of the twins was deemed impossible. Since the parents desired survival of one twin, if possible, the institution's ethics committee met, and they recommended surgical intervention. A separation procedure was designed for baby A, the only infant thought to be salvageable.

Two days prior to surgery, a plan for perioperative management was designed by consultation between the anesthesiologists, pediatric cardiac surgeon, pediatric surgeon, pediatric cardiologist, neonatologists, pathologist, and the operating room staff. Though no rehearsal of the operative procedure was undertaken prior to the separation, requirements for each of the individuals participating were communicated.

Separation Procedure. The twins were transported to the operating room supported by asynchronous positive-pressure ventilation. Two teams of anesthesiologists participated, each consisting of staff and resident members. A cardiopulmonary technician was available to both teams. Ventilation with an air–oxygen mixture ($F_{I_{O_2}}$ 0.5) was established asynchronously with time-cycled, pressure-limited ventilators (BABYbird®). Esophageal electrocardiographic monitoring was employed in baby A as previously described[13] because of limited available skin surface for lead placement.

Anesthesia was induced over a 5-min period with fentanyl, 100 μg/kg iv and paralysis with pancuronium, 0.15 mg/kg iv. No additional fentanyl was administered throughout the case. Supplemental doses of pancuronium provided muscular relaxation. A left radial arterial catheter and a central venous catheter (facial vein cutdown) were inserted into baby A. Baby B underwent only central venous catheter placement. Rectal temperature was monitored continuously in baby A. With the infants rotated 180 degrees about the craniocaudad axis, antiseptic preparation of the eventual dependent skin surface was accomplished. They then were placed upon a prepared sterile operating room table and their superior surface antiseptically prepared.

A continuous dopamine infusion, at 3.0 $\mu g \cdot kg^{-1} \cdot min^{-1}$, used to theoretically improve renal blood flow, was begun prior to the initial

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* Staff Anesthesiologist.
† Anesthesiology Resident.
‡ Cardiothoracic Surgeon.

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Address reprint requests to Dr. Brown: Department of Anesthesiology, The Mason Clinic, Box 900, Seattle, Washington 98111.

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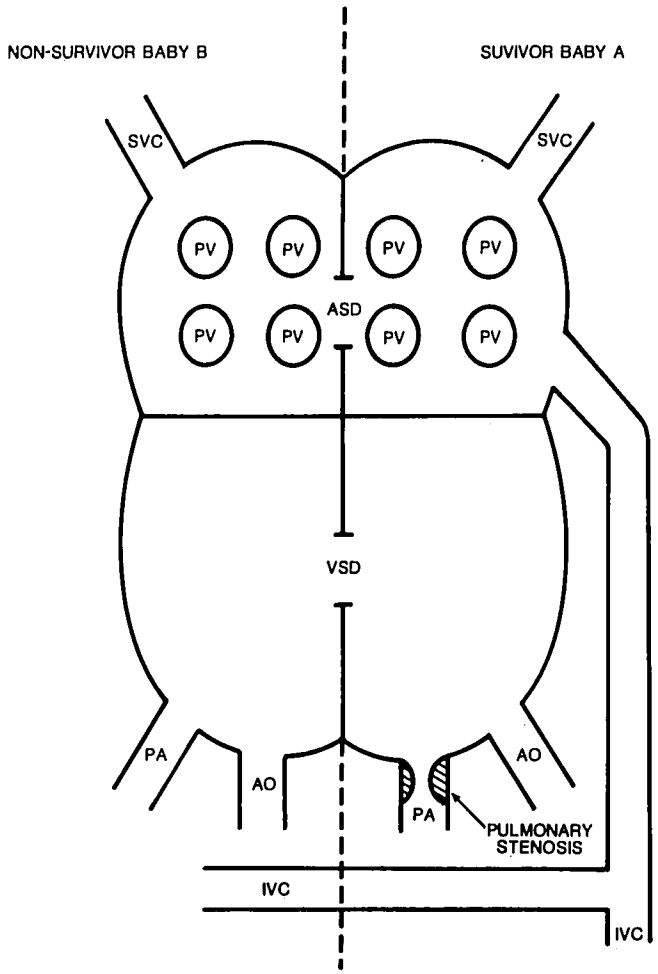

FIG. 1. Cardiac anatomy of thoracopagus, cardiopagus twins prior to separation. SVC = superior vena cava; IVC = inferior vena cava; ASD = atrial septal defect; VSD = ventricular septal defect; AO = aorta; PA = pulmonary artery; PV = pulmonary vein.

surgical procedure, an exploratory thoracotomy. During this procedure a cardiac arrest occurred, which was thought due to atrial distention following transfusion of packed erythrocytes (15 ml) at a time when blood loss was being replaced milliliter for milliliter. The twins were resuscitated rapidly with epinephrine, atropine, and withdrawal of 20 ml blood through the central venous pressure (CVP) catheter. Immediately prior to the arrest, Pa_{O_2} was 58 mmHg, Pa_{CO_2} 37 mmHg, *p*H 7.39 units, H_{CO_3} 23.5 mEq/l. An exploratory laparotomy then was performed and revealed shared small bowel and liver.

The aorta of baby B, arising from its topographic right ventricle, was used to bypass the stenotic pulmonary artery of baby A. Baby B died when its aorta was divided. However, during baby A's aorta-to-pulmonary shunt anastamosis, oxygenation was provided via the pulmonary circulation and lungs of baby B. When the shunt anastamosis was completed, baby B's pulmonary artery, superior and inferior vena cavae, and pulmonary veins were ligated. The cardiovascular anatomy of baby A then was as illustrated in figure 2.

Following completion of the cardiac portion of the procedure, division of the common liver was undertaken. Hemorrhage of an estimated 325 ml (1.3 times the estimated blood volume) occurred requiring blood and fluid administration as outlined in figure 3. Subsequent attempted closure of the chest and abdomen led to cardiac tamponade and ventricular fibrillation. Successful resuscitation followed treatment with epinephrine, lidocaine, and dc defibrillation (5-watt seconds). The diaphragm was left divided to forestall further tamponade. Subsequent skin closure incorporated silastic-reinforced Dacron® sheeting for both chest and abdomen to form a "chimney," which was to be closed incrementally. Baby A then was transported to the NICU in satisfactory condition. On admission, arterial blood pressure was 50/35 mmHg, heart rate 165 bpm, pH_a 7.45, Pa_{O_2} 80 mmHg, Pa_{CO_2} 29 mmHg, and hemoglobin 12 g/dl.

On the first postoperative day, supraventricular tachycardia was corrected by overdrive pacing after placement of epicardial leads. On the 9th postoperative day the infant underwent final closure of the diaphragm and abdominal wall. Diaphragmatic closure was accomplished in part by utilization of a bovine pericardial patch. At the time of reoperation, a small duodenal perforation was noted incidentally and oversewn. This was thought to represent necrosis, secondary to high intraabdominal pressures generated from progressive, daily chest and abdominal closure of the silastic chimney.

The infant survived with ventilatory support for 2 months following

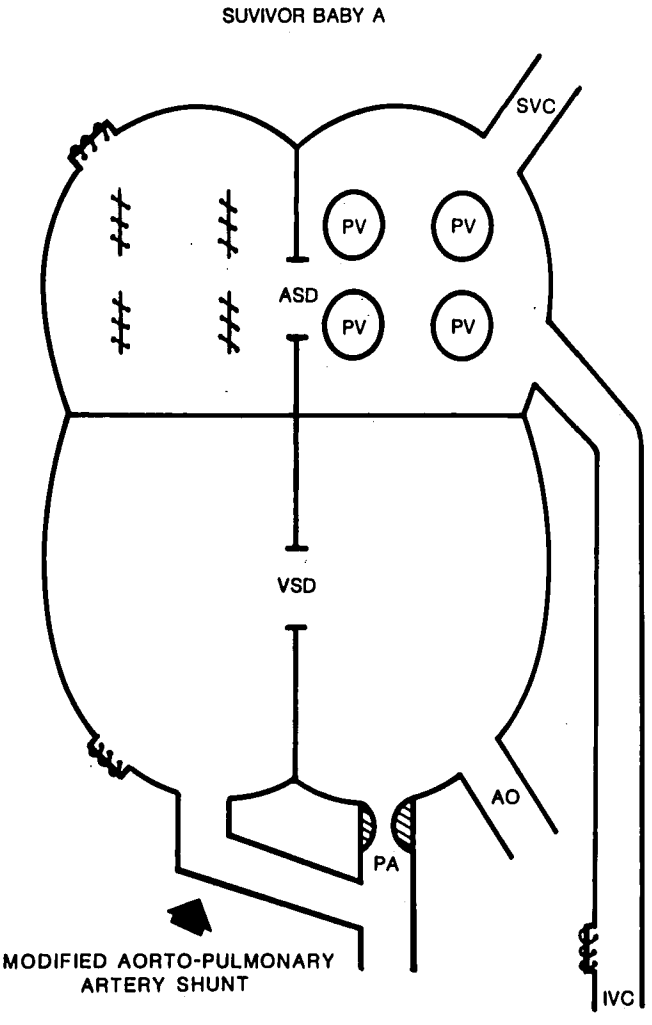

FIG. 2. Cardiac anatomy of survivor, baby A, after separation. SVC = superior vena cava; IVC = inferior vena cava; ASD = atrial septal defect; VSD = ventricular septal defect; AO = aorta; PA = pulmonary artery; PV = pulmonary vein.

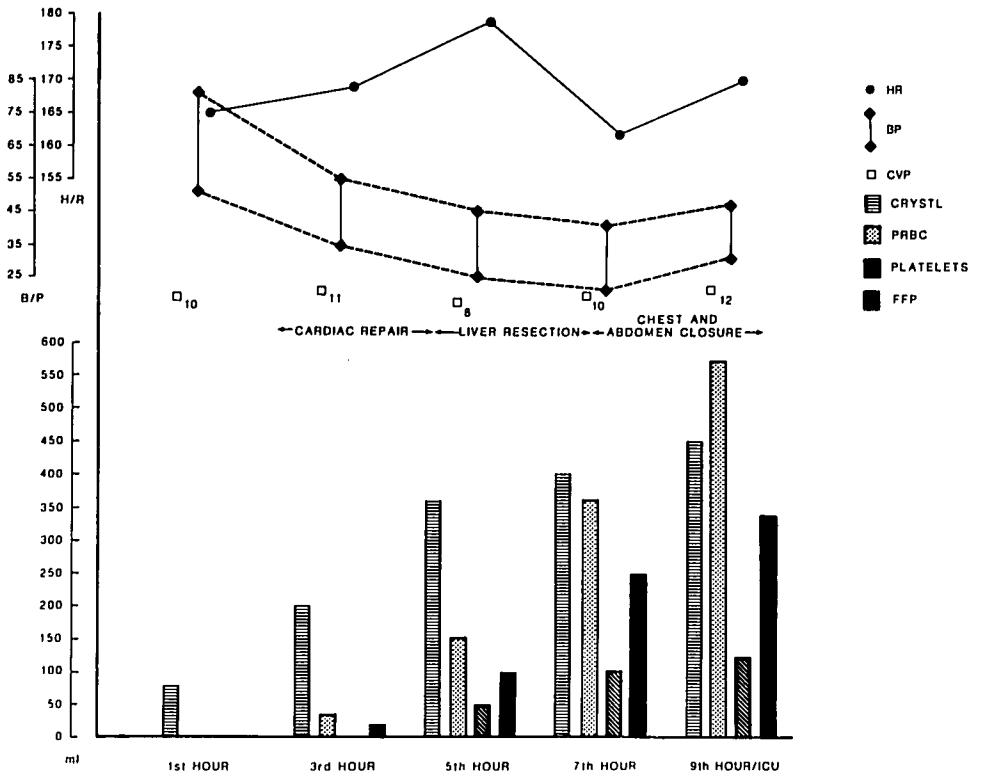

FIG. 3. Cumulative summary of fluid and blood administration and hemodynamics throughout operative procedure. HR in beats/min; BP in mmHg; CVP in mmHg.

the second procedure but eventually succumbed to progressive heart failure. Multiple echocardiograms did not identify a correctable cardiac lesion. At postmortem the superior vena cava and the modified aortopulmonary artery shunt were obliterated by thrombus. Superior vena cava obstruction was suspected but unproven antemortum secondary to generalized peripheral edema. Cardiac catheterization was not performed, since additional surgical intervention was considered to impose unacceptable risk.

DISCUSSION

Survivors from separation of cardiopagus–thoracopagus twins have been those twins sharing only atrial tissue.[12] The lesion we described has been uniformly lethal whether operated upon or not. Management of this separation created special concerns involving the perioperative anesthetic and cardiovascular management.

Preoperatively, our concerns were centered upon ethical issues, cardiovascular anatomy, procedural rehearsal, and timing of operative repair. Only after the parents and the institution's ethics committee contemplated the issues and urged an attempt to allow one infant a chance at survival was surgical separation considered. All individuals who would care for the twins in the immediate perioperative period were asked to examine their feelings regarding appropriateness of the intervention and to excuse themselves if conflict arose.

Cardiovascular anatomy clearly was outlined preoperatively and allowed planning for the use of baby B's lungs as an *in vivo* oxygenator while the aortopulmonary artery shunt was created in baby A. This technique has not been reported previously. The procedural rehearsal was conducted in a detailed verbal fashion rather than as an operating room event. This fact did not indicate superiority of our method over prior recommendations,[12] but rather an alternative approach. Timing of separation also was considered preoperatively. We decided to separate the twins prior to cardiac decompensation and the complications of multiorgan failure.

Contemplated intraoperative problems included operating room crowding, choice of anesthetic technique, iv fluid management, cardiovascular monitoring, infant temperature preservation, and closure of the surviving twin. The procedure was scheduled for a Sunday to minimize operating room congestion, and only members of the operating team were allowed in the immediate area. Traffic flow was monitored by an operating room nurse. Questions regarding acute patient management were directed only to the primary operating surgeon and anesthesiologist. To conserve space, the anesthesia machine was moved out of the operating room suite and ventilation provided by two infant ventilators. Because of free mixing of the infant's circulations, we elected to anesthetize the twins with a single injection of fentanyl, 100 μg/kg.[14] The anesthesiologists then could concentrate on iv fluid management, which was of primary importance throughout the procedure. Arterial cannulation was necessary to monitor arterial

blood pressure as well as to allow frequent sampling of arterial blood for measurement of gas tensions and *p*H. We considered, as did Bloch and Karis,[12] that CVP catheters were essential to guide iv fluid therapy, as well as to provide central circulatory access in the event cardiac resuscitation was required. We employed a 5-French double-lumen CVP catheter, designed for pediatric use by the senior author, which allowed continuous measurement of central venous pressure during simultaneous transfusion and drug therapy.

In contrast to prior recommendations,[12] most of our monitoring devices were placed in the operating room in order to simplify transport. Previous investigators suggested the monitors be placed in the intensive care nursery to conserve the twins' temperature. The temperature of the surviving infant (baby A) did not fall below 37° C any time during the preparatory phase and was 36.8° C when she was transported to the NICU at completion of the procedure. Heat loss was minimized by keeping room temperature above 28° C and by the utilization of a heat lamp intermittently directed upon the infant. In addition, all fluid and blood products were warmed with a similar heat lamp prior to infusion. The potential problem of cardiac arrest associated with skin closure had been anticipated preoperatively. Although primary skin closure initially appeared possible, it was not. This emphasizes again that closure is a critical step in such procedures.

Postoperative problems that we considered included the transition from operating room care to NICU and potential renal dysfunction secondary to heart failure. The anesthesiologists continued to manage the iv fluid and cardiovascular function for 30 min after stabilization in the NICU to minimize the hemodynamic change associated with transfer. We utilized "dopaminergic" levels, 3 $\mu g \cdot kg^{-1} \cdot min^{-1}$, of dopamine for its theoretic benefit on renal blood flow. Though this use of dopamine has not been documented in pediatric patients, it is well established in adult cardiac surgical patients.[15] Intraoperative placement of pacing wires was considered. Retrospectively, this procedure would have simplified postoperative management. The recognition of superior vena caval thrombosis at autopsy suggests that central venous catheters be removed as soon as possible, though the more likely etiology for thrombosis was altered cardiovascular anatomy.

In summary, we describe the perioperative course of the longest separated survivor of cardiopagus twins joined at both atria and ventricles. The initial success of this procedure is attributed to clear communication between members of the separation team, accurate preoperative understanding of the cardiovascular anatomy, and careful iv blood and fluid replacement.

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