Separation of Conjoined Twins Utilizing Cardiopulmonary Bypass

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Abstract

The incidence of conjoined twins occurs once in every two million live births. Successful separation has occurred in 50% of these patients. Previous attempts to separate twins at the sagittal sinus have resulted in death. On September 5, 1987, the team at Johns Hopkins Hospital was the first to successfully separate conjoined twins at the sagittal sinus. One key to this success was the use of cardiopulmonary bypass, deep hypothermia and circulatory arrest.

Each patient was cannulated with a 14 Fr. aortic cannula and a 22 Fr. venous cannula in the right atrium. They were then connected to completely separate cardiopulmonary bypass circuits. Cardiopulmonary bypass was initiated utilizing a roller pump, pediatric membrane oxygenator with integral cardiotomy, and an arterial filter. The twins were cooled to 20 degrees centigrade in preparation for circulatory arrest. During circulatory arrest, separation occurred and repair was accomplished utilizing direct suture and pericardial patches. When repair was complete, cardiopulmonary bypass was resumed and rewarming begun. At a rectal temperature of 33 degrees centigrade, cardiopulmonary bypass was discontinued.

The two major benefits of extra-corporeal circulation in this procedure were maintaining hemodynamic stability and the ability to regulate metabolic demands. The use of cardiopulmonary bypass was a solution to the obstacles presented. The ability to control blood flow, temperature and volume resulted in a successful separation of these patients.

Introduction

The incidence of conjoined twins occurs once in every two million live births[1]. This case study will deal with the separation of a pair of conjoined males, patients who were joined at the superior sagittal sinus. It is one of the sinuses of the dura mater (Figure 1). The sagittal sinus is lined with endothelial tissue which is continuous with the lining of the circulatory system.[2] It is one of five sinuses located in the postero-superior section of the skull.[3]

The superior sagittal sinus receives the superior cerebral veins, veins from the diploe and dura mater, and near the posterior extremity of the sagittal suture, veins from the pericranium, which pass through the parietal foramina.[4]

Due to the extensive communication and the large amount of venous blood return through the sagittal sinus, it was apparent that the separation and reconstruction necessary would make it technically extremely difficult to divide the sinus without damage to the brain and the possibility of perioperative death. It was with these concerns in mind that the use of cardiopulmonary bypass was investigated. Deep hypothermia and circulatory arrest have been employed to

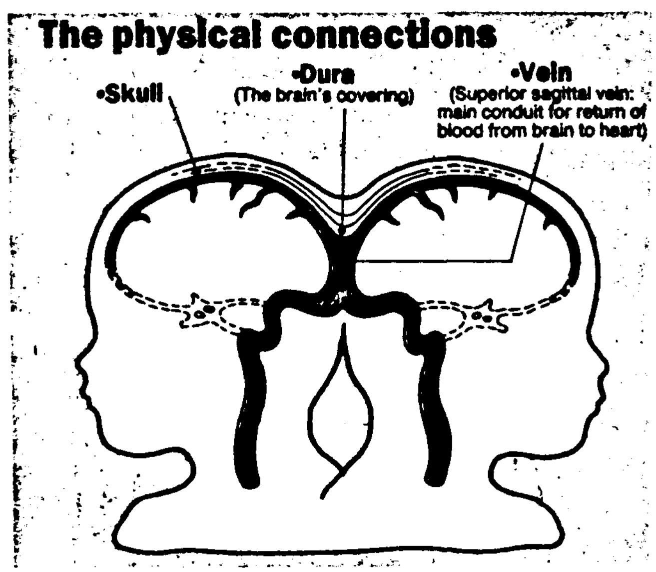

Figure 1: Demonstration of the communication of the superior sagittal sinus vein, (reprint by permission of the Baltimore Sun for AmSECT).

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provide hemodynamic stability, while regulating metabolic demands.[5] The utilization of circulatory arrest during neurosurgical procedures would provide the neurosurgery team with a bloodless field in which to work for approximately one hour. It was felt that the necessary repair could be completed in this period.

The use of cardiopulmonary bypass with deep hypothermia and circulatory arrest has many associated risks.[6] The greatest source of morbidity with this procedure would be bleeding. Intraoperative bleeding would be exacerbated by heparinization, mechanical coagulopathies arising from blood interaction with foreign surfaces. These include decreased platelet numbers and function and an increase in red blood cell fragility. Deep hypothermia and circulatory arrest present other problems such as tissue edema due to circulatory standstill and the time limitations. The length of time on cardiopulmonary bypass necessary to rewarm can also be a detrimental aspect of the procedure. Although these were potential problems, cardiopulmonary bypass with deep hypothermia and circulatory arrest was viewed as a vital adjunct to this procedure that would allow a successful outcome.

With the procedure mapped out, the operating room needed to be arranged to accommodate the demands of this surgical procedure. The surgery suite had to be reconstructed with a duplication of all equipment. The desired result was to create a room that was a mirror image of itself, able to handle two simultaneous procedures on two separate people (Figure 2).

Materials and Methods

Two complete teams, one for each child, were assembled for the separation of this set of conjoined twins. The combined teams consisted of 12 surgeons (5 neuro, 2 cardiac, 3 plastic, 2 oral), 8 anesthesiologists, 20 nurses, 5 cardiopulmonary technicians, 3 perfusionists, 6 nursing technicians, 5 electrical engineers, 2 photographers, 4 audiovisual personnel, and 4 security personnel. The operation required duplication of all instrumentation including 2 cardiopul-

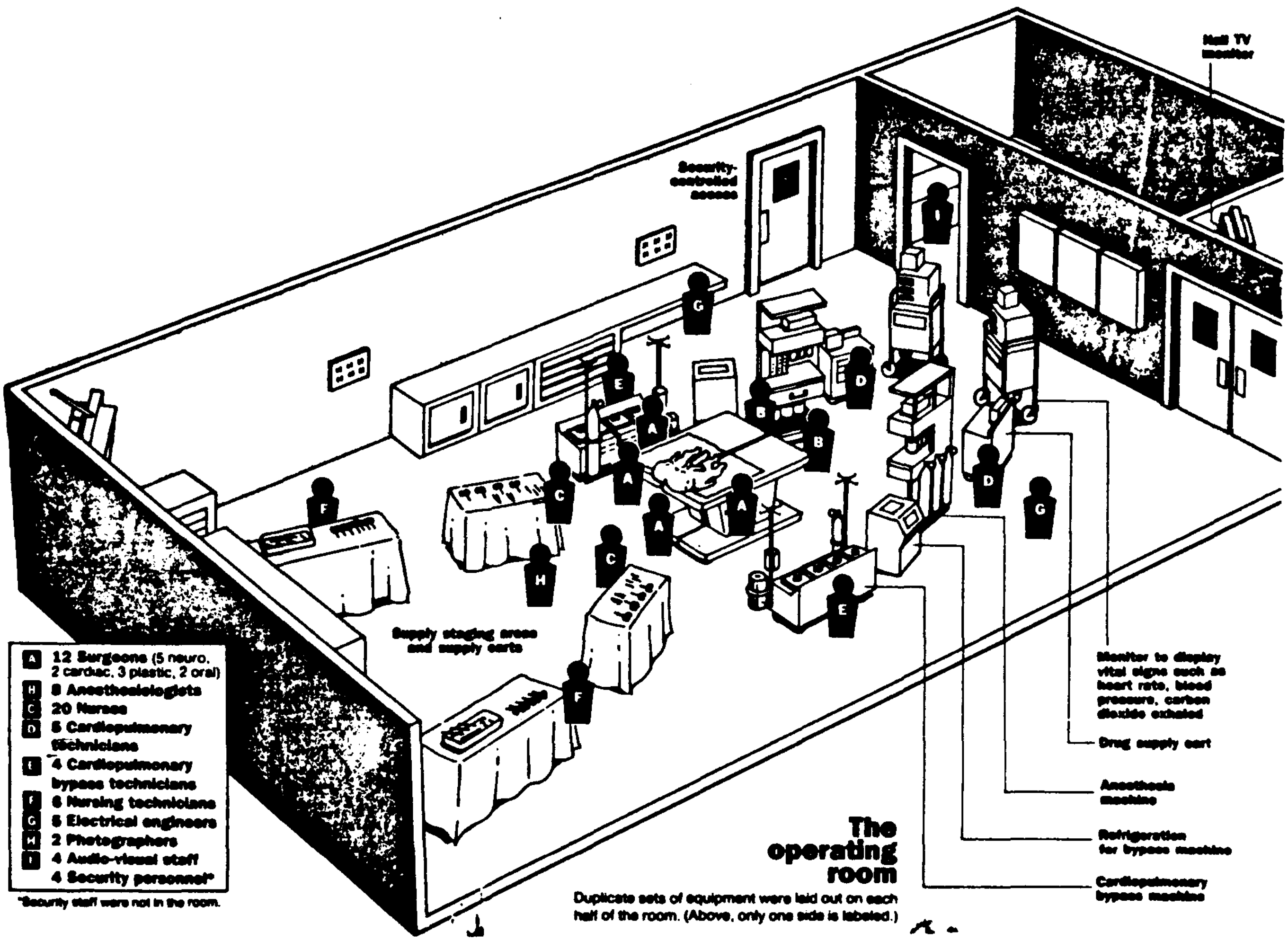

Figure 2: Graphic illustration of the operating room and associated personnel (reprint by permission of the Baltimore Sun for AmSECT).

monary bypass circuits. The pump circuits consisted of a membrane oxygenator with integral cardiotomy, Cobe VPCML[a], arteriovenous tubing (¼″), and arterial filter, Pall LPE 1440.[b] Continuous inline arterial and venous blood gas monitoring using a CDI Model 300[c] was also employed. The patients were brought into the operating room and placed on adjoining operating room tables. The twins were placed under general anesthesia with endotracheal ventilation. Routine invasive monitoring lines were then inserted by the anesthesia staff.

Neurosurgeons then proceeded to dissect down to the level of the common sagittal vein. Following this, cardiac surgeons operating on each child independently, performed sternotomy and cannulated for cardiopulmonary bypass. Each aorta was cannulated with a 14 Fr. aortic cannula and the right atrium with a 22 Fr. venous cannula.

During cannulation, the bypass circuits were primed. The prime consisted of one unit whole blood, that had been heparinized with 2000 u beef lung heparin, recalcified with 500 mg CaCl, and one unit of fresh frozen plasma. Total added volume was 700 cc. Mannitol (0.5 gm/kg) and a calculated dose of sodium bicarbonate was added to the prime.

Each patient was heparinized according to a 300 u/kg of beef lung heparin protocol. Extension of the ACT was verified for each child. Simultaneously cardiopulmonary bypass was initiated and flows were increased until a rate of 100 ml/kg was achieved. Patient cooling was carried out concurrently. Flow was decreased to 50 ml/kg, when the rectal temperature reached 20 degrees centigrade. During the cooling period neurosurgeons continued to dissect down to the level of the sagittal vein. At this point, flows were decreased to 20 ml/kg at a rectal temperature of 17 degrees centigrade. The aortas were then cross-clamped. A cardioplegia dose of 10 ml/kg was administered to arrest the heart. With the hearts arrested the pumps were simultaneously shut off and complete exsanguination was allowed to take place. The cannulas were clamped and disconnected. The arterial and venous tubing were then reconnected to each other, and the blood allowed to recirculate.

During cardiopulmonary bypass, acid base balance and venous oxygen saturation were monitored using an inline blood gas monitor. These results were compared and correlated with values from the hospital laboratory. Arterial line pressures were also monitored.

When circulation was arrested, neurosurgeons separated the common superior sagittal sinus and the operating room tables were moved apart. The repair of each child was then carried out by two separate teams working simultaneously. Although similar procedures were planned for both children, it became evident that some differences in repair were necessary. One patient's sinus was repaired with a patch made from pericardium (Figure 3), and the other patient was a direct repair. The repair took 63 minutes for Baby A and 52 minutes for Baby B. As the repair was completed each patient was reconnected to their respective cardiopulmonary bypass circuits. Circulation was restored with simultaneous rewarming. At rectal temperatures of 33 degrees centigrade cardiopulmonary bypass was discontinued. Following

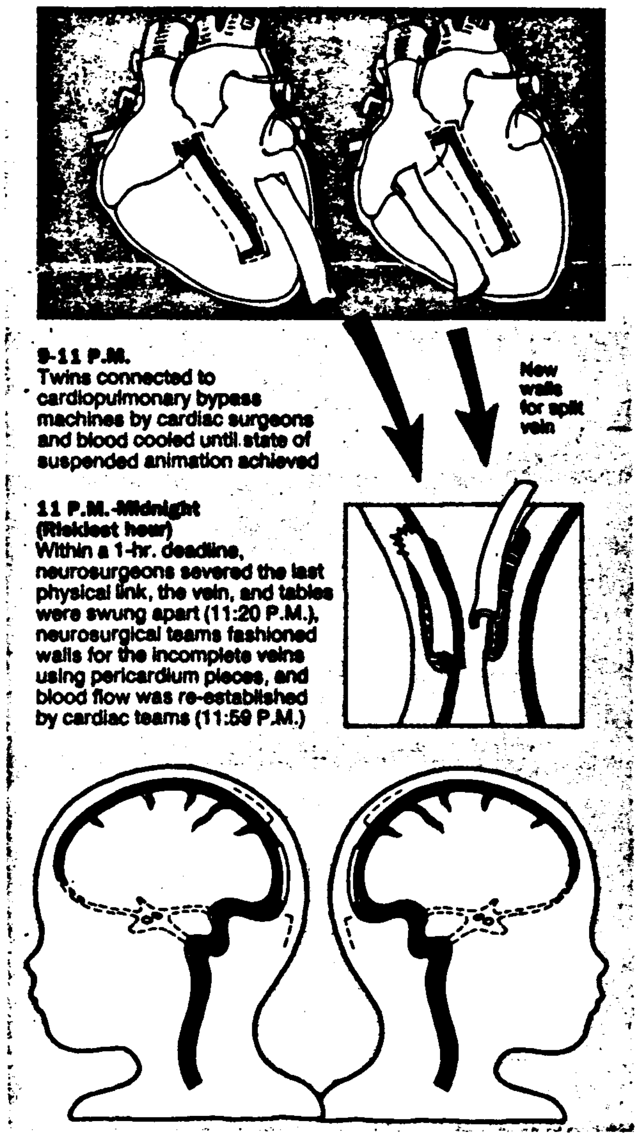

Figure 3: Demonstration of the pericardial patch placement (reprint by permission of the Baltimore Sun for AmSECT).

a Cobe Laboratories, Lakewood, CO 80215
b Pall Corporation, Glen Cove, NY 11542
c Cardiovascular Devices Inc., Irvine, CA 92714

decannulation, heparin reversal and hemostasis was accomplished. The repair of the sagittal sinus continued and reconstruction was completed.

Results

Perioperative hemorrhage was more extensive than anticipated. With the brain totally exposed, general oozing progressed to extensive bleeding. The communication of the sagittal sinus was greater than anticipated. During the initial phases of rewarming, neurosurgeons repaired sinus and dural bleeding sites with cautery and thrombin. Topical thrombin is a commonly used hemostatic medium in neurosurgery. Hemorrhage was severe enough to decrease venous drainage to near zero. This situation required continous transfusion of Lactated Ringers and packed red blood cells. During this period it was discovered that the neurosurgeons were allowing large amounts of blood to drain from each child's wound. This blood was being collected in basins and not communicated to the other members of the team. This shed blood was discarded as it contained thrombin. Prior to the surgical procedure, it was decided that an operative field cell salvage system would not be used due to the small blood volumes involved. When hemorrhaging persisted, a Cell Saver 4[d] was employed. Nevertheless, thrombin was returned to the pump circuit requiring one oxygenator to be changed. Surgeons were notified, the oxygenator changed, and cardiopulmonary bypass was resumed without incident. The other oxygenator developed clots in its venous reservoir but did not require replacement.

Changes in the systemic vascular resistance between Baby A and Baby B prior to separation resulted in an unexpected shunting of blood via the sagittal sinus. Baby A had a low systemic vascular resistance and mean perfusion pressures of 20–25 mmHg at flows of 100 ml/kg/min. Baby B maintained a mean perfusion pressure of 40 mmHg at similar flows. As hypothermia deepened, the systemic pressure of Baby A increased to 40 mmHg. The correction of the pressure gradient between the patients alleviated the shunting of venous blood through the sagittal sinus. Shunted blood was removed from Baby A's cardiopulmonary bypass circuit and transfused into Baby B's cardiopulmonary bypass circuit using empty plasma transfer bags with a capacity of 500 ml. It was noted that Baby A's oxygenator was about two inches lower than Baby B's; this was corrected and venous drainage was equalized.

d Haemonetics Corporation, Braintree, MA 08216

Discussion

Despite practice sessions and meetings prior to surgery, which were held in an attempt to simulate any problems, there were complications which arose. Due to the anatomy, once separated, Baby A received direct suturing of the sagittal sinus and Baby B received a pericardial patch. The venous communication was significant enough to cause massive shunting between Baby A and Baby B when pressures were not equal. But most importantly, the blood loss from the patient's heads was a greater than expected complication. The use of thrombin prohibited shed blood from being returned to the pump circuit. Venous return was poor necessitating the transfusion of large amounts of red blood cells and Lactated Ringers.

Constant monitoring of arterial blood gases by an inline monitor showed values which were grossly abnormal. Later it was realized that the cardiopulmonary bypass circuit was acting as a rapid infusion system. The arterial blood gas values were essentially an analysis of the red blood cells and Lactated Ringers infused, not the patient's true acid base status. Once bleeding was controlled venous return was adequate to maintain flows of 100 ml/kg. Arterial blood gases were corrected, warming proceeded and cardiopulmonary bypass terminated.

The major benefits of extra-corporeal circulation with deep hypothermia and circulatory arrest in this procedure were maintenance of hemodynamic stability and the ability to reduce metabolic demands. The period of circulatory arrest provided a bloodless field for the surgical repair. Along with these benefits, there are definite problems associated with the use of cardiopulmonary bypass. These risks include, bleeding due to heparinization, decreased platelets number and function, as well as other mechanical coagulopathies. Other limitations of deep hypothermia include the time on bypass required for cooling and rewarming and increased edema due to circulatory arrest. The circulatory arrest period is restricted to approximately one hour. With all of this in mind, the use of cardiopulmonary bypass was the solution to the problems presented. The ability to control temperature and volume resulted in a successful separation of these patients.

In retrospect, if this situation should occur again there are some things that could be done differently. More communication between team members is needed as visibility was a problem. Cell salvage should be on standby and the blood bank should be better prepared to provide increased amounts of red blood cells. Thrombin should be used cautiously during cardiopulmonary bypass.

Baby A and Baby B have had a long and tough course. They are now preparing to go home after several months at The Johns Hopkins Hospital. At one year of age, and six months after separation, they are recovering satisfactorily.

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