in preparation for a Norwood procedure. This approach proved to be a simple, quick, and remarkably effective means of restoring a satisfactory systemic perfusion, providing the appropriate circulatory conditions for the recovery of multiple organ dysfunction in these 2 critically ill neonates before a Norwood procedure in the newborn period.

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CASE REPORTS

Pulmonary-to-Systemic Blood Flow Ratio Oriented Management After Repair of Obstructive Total Anomalous Pulmonary Venous Connection in Neonates With Single Ventricle

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Caval oxygen saturation was monitored to estimate pulmonary-to-systemic blood flow ratio after relief of obstructive total anomalous pulmonary venous connection in two neonates with single ventricle. Distribution between systemic and pulmonary blood flow was manipulated by pharmacologic, ventilatory, and surgical interventions aimed at achieving pulmonary-to-systemic blood flow ratio of 0.5 to 1.0. Monitoring of pulmonary-to-systemic blood flow ratio facilitates appropriate balancing between tissue perfusion and oxygenation, and detects redundant ventricular volume-load.

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Recent articles indicate the usefulness of mixed venous oxygen saturation (SvO_2) monitoring to estimate pulmonary-to-systemic blood flow ratio (Qp/Qs) in perioperative management of the Norwood procedure [1,2]. We monitored Qp/Qs after repair of pulmonary venous obstruction in two neonates with right isomerism, single ventricle (SV), and supracardiac total anomalous pulmonary venous connection (TAPVC).

Patient 1, a previously asymptomatic female infant, exhibited dyspnea at 20 days of age. She was taken to a community hospital in an ambulance. Because of profound cyanosis, she was intubated, given alprostadil (PGE1), and transferred to our hospital. On admission, arterial blood gas analysis revealed arterial oxygen saturation of 0.47 and acid-base balance of −4.9. Chest radiograph revealed dextrocardia, normal heart size, bilaterally right-sided bronchial arrangement, ground-glass like lung fields, and central liver. Echocardiography disclosed left-sided aortocaval juxtaposition, bilateral superior vena cava (SVC), obstructive total anomalous pulmonary venous connection to the left-sided SVC, single atrium, unbalanced complete atrioventricular septal defect with dominant right ventricle, ventricular l-loop, d-malposition of the great arteries, and pulmonary stenosis. After resuscitation and stabilization, operation was performed the next morning. Intraoperative inspection revealed isomeric right atrial appendages and vertical vein obstruction at its junction with the left-sided SVC. She underwent common pulmonary vein-to-common atrium anastomosis. The vertical vein was left patent. After surgery, the sternum was left open. Qp/Qs was calculated from arterial oxygen saturation (SaO_2) and right-sided SVC oxygen saturation as a surrogate of SvO_2, assuming pulmonary venous oxygen saturation to be 0.96 (Fig 1). Qp/Qs gradually increased from 1.33 before skin closure to 2.9 at 55th postoperative hour despite pharmacologic and ventilatory manipulations to decrease Qp/Qs. Therefore, pulmonary artery banding was performed concomitantly with sternal closure. Qp/Qs after pulmonary artery banding ranged between 0.51 and 1.29. Endotracheal tube was removed at 110th postoperative hour. Staged total cavopulmonary connection was completed at 3 years old. At 4 years old, although she suffers from neurologic disorder, her cardiopulmonary condition is stable.

Patient 2, a 1-day-old male infant exhibiting cyanosis and tachypnea, was transferred from an obstetrician. Arterial blood gas analysis in room air revealed oxygen saturation of 0.6 and normal acid-base balance. Chest radiograph revealed levocardia, normal heart size, bilaterally right-sided bronchial arrangement, subtly hazy lung field, central liver, and left-sided gastric bubbles. Echocardiography revealed right-sided aortocaval juxtaposition, total anomalous pulmonary venous connection to the left innominate vein, unbalanced complete atrioventricular septal defect with dominant right ventricle, ventricular d-loop, d-malposition of the great arteries, pulmonary atresia, and patent ductus arteriosus. Pulsed

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Doppler study revealed pressure gradient across vertical vein obstruction of 16 mm Hg. He was given oxygen, but not intubated. Alprostadil was administered. His stable condition predisposed us to postpone surgery until 30 days of age. At surgery, right isomerism of the atrial appendages was confirmed. The vertical vein passing behind the left-sided pulmonary artery was compressed by a vise formed by the left-sided pulmonary artery and left-sided bronchus. Common pulmonary vein-to-common atrium anastomosis, ligation of patent ductus arteriosus, and placement of a 3.5-mm prosthetic innominate artery-to-pulmonary artery shunt were performed. The vertical vein was left patent. Venous blood sampling line was inserted in the inferior vena cava. SaO_2 and Qp/Qs before sternal closure were 0.52 and 0.50, respectively (Fig 2). After surgery, dobutamine, isoproterenol, and nitric oxide were administered to increase cardiac output and Qp/Qs. After Qp/Qs exceeded 1.0 at the second postoperative hour, dobutamine, isoproterenol, and nitric oxide were tapered to counteract the intrinsic tendency of increasing Qp/Qs. Endotracheal tube was removed at 61st postoperative hour. At 6 months of age, he waits for staged Fontan operation at home.

Comment

Appropriate distribution between systemic and pulmonary blood flow is essential after obstructive TAPVC repair in neonates with SV: diminished Qp/Qs causes hypoxemia, whereas excess Qp/Qs causes insufficient tissue perfusion. Previous reports documented increasing pulmonary blood flow and resultant prolonged ventricular volume overload frequently leading to fatal heart failure after TAPVC repair in neonates with SV [3].

Perioperative management would have been different if we had not used Qp/Qs monitoring. In patient 1, if we had not been aware of high Qp/Qs before delayed sternal closure, SaO_2 of 0.8 without signs of heart failure would have warranted sternal closure without pulmonary artery banding. In patient 2, if we had not known Qp/Qs to be within acceptable range after cardiopulmonary bypass, low SaO_2 would have urged us to revise systemic-to-pulmonary shunt. Although Qp/Qs fluctuated considerably, it was regulated within a tolerable range in our patients. Reportedly, stable Qp/Qs was maintained after the Norwood procedure with the use of Qp/Qs monitoring [4]. Qp/Qs monitoring facilitates appropriate balance between tissue perfusion and oxygenation, as well as early detection of redundant ventricular volume load before signs of excessive pulmonary flow or heart failure develop.

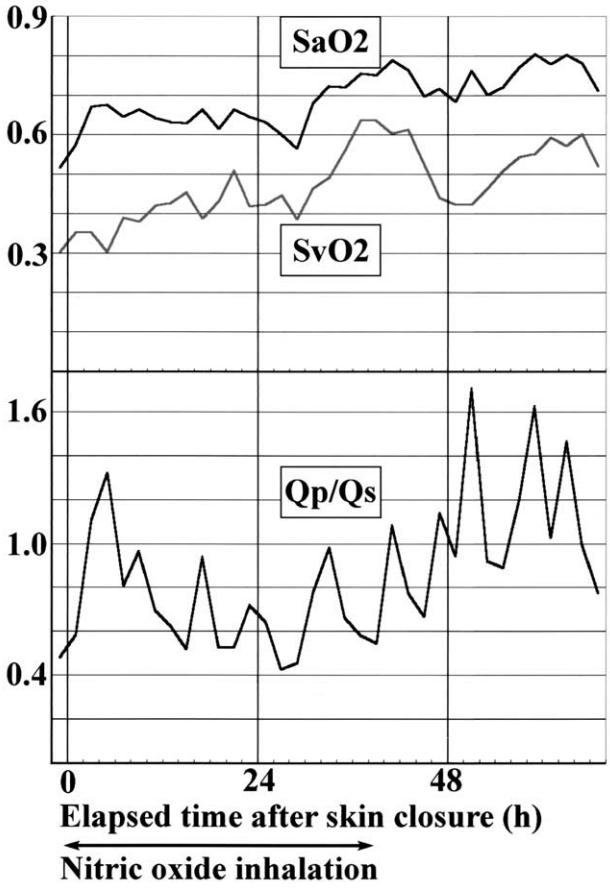

Fig 2. Perioperative changes in arterial oxygen saturation (SaO2), superior vena cava oxygen saturation (SvO2), and pulmonary-to-systemic blood flow ratio (Qp/Qs) in patient 2. Qp/Qs was initially low, but gradually increased in the first 48 hours.

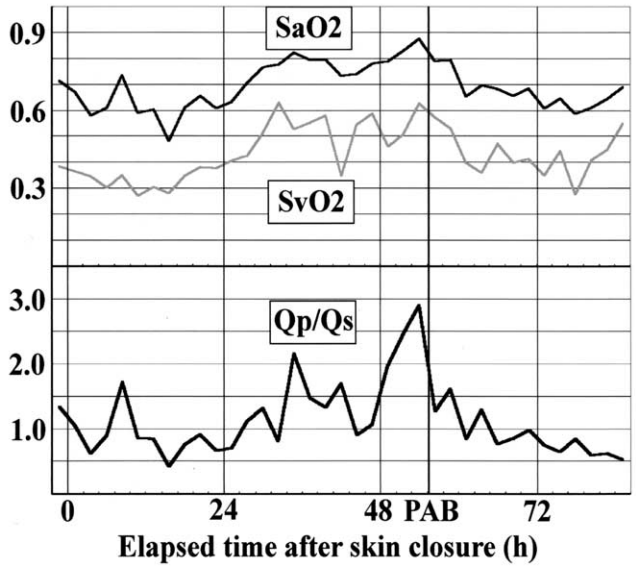

Fig 1. Perioperative changes in arterial oxygen saturation (SaO2), superior vena cava oxygen saturation (SvO2) and pulmonary-to-systemic blood flow ratio (Qp/Qs) in patient 1. SaO2 and SvO2 appeared favorable before pulmonary artery banding (PAB), but calculated Qp/Qs was unacceptable.

Target range of Qp/Qs in neonate with SV has not been elucidated. Barnea and colleagues [5] suggested that maximal oxygen delivery is attained with Qp/Qs of 0.5 to 1.0. Tweddell and associates [4] stated that their postoperative management after the Norwood procedure was aimed at achieving Qp/Qs of 0.8 to 1.2. Pearl and associates [6] indicated that optimal Qp/Qs is in the range of 0.7 to 1.0. Our policy is the following: when anaerobic metabolism is indicated by depleted SvO_2 as low as 0.3, accumulated blood lactate, and metabolic acidosis, then Qp/Qs is targeted at 1.0 to obtain maximal tissue oxygen saturation because mathematical model predicts that Qp/Qs of 1 provides maximal SvO_2 [2]; when SvO_2, lactate, and acid-base balance indicate aerobic metabolism, then Qp/Qs is targeted between 0.5 and 1.0 to prepare for increasing Qp/Qs.

Possible drawbacks of Qp/Qs-oriented management in neonates with TAPVC and SV are twofold. First, placement of caval blood sampling line or spectrophotometric catheter is invasive. It may cause infection or thrombosis. Frequent blood sampling increases transfusion dose, and thereby chance of its adverse effects. Second, calculated Qp/Qs may deviate from the true value. Pulmonary congestion or atelectasis may make pulmonary venous oxygen saturation below 0.96. Placement of caval catheter close to the atrium or in the cava connected to the pulmonary veins allows pulmonary venous blood contamination in venous samples, making measured caval oxygen saturation deviate higher. If pulmonary venous oxygen saturation is below 0.96 or caval oxygen saturation is incorrectly high, calculated Qp/Qs can be lower than the true value, leading to incorrect patient management. Nevertheless, we believe that Qp/Qs monitoring facilitates appropriate balancing between tissue perfusion and oxygenation, detects redundant ventricular volume-load, and will improve outcome of obstructive TAPVC repair in neonates with SV.

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CASE REPORTS

Anatomic Correction for Corrected Transposition After Pulmonary Unifocalization

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A Senning plus Rastelli operation was performed in a patient who had a rare combination of congenitally corrected transposition of the great arteries (S,L,L) with dextrocardia, major aortopulmonary collaterals, and diminutive central pulmonary arteries with arborization defects. The patient required four preparatory operations including bilateral unifocalizations of the aortopulmonary artery collaterals. Pulmonary artery to systemic pressure ratio after the double switch operation was 0.6. The patient demonstrates good biventricular function on echocardiogram at 3 months after the operation.

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The use of the double switch operation (DSO) for the management of congenitally corrected transposition of the great arteries (ccTGA) is becoming increasingly widespread [1]. Surgical management is especially difficult in the presence of diminutive central pulmonary arteries with arborization defects and major aortopulmonary collateral arteries (MAPCAs). We encountered this very rare disease entity, which required staged unifocalization of the MAPCAs before the DSO.

An 8-month-old male patient was referred with ccTGA (S,L,L) with a large ventricular septal defect, atrial septal defect, dextrocardia, pulmonary atresia with very small confluent pulmonary arteries and arborization defects. A total of five MAPCAs supplied nine pulmonary segments in the following pattern: two segments of the right upper lobe, three segments of the right lower lobe, two segments of the left lower lobe, and two segments of the left upper lobe. The central pulmonary arteries measured 1.5 to 2 mm on angiogram. Echocardiography demonstrated normal biventricular function without tricuspid or mitral regurgitation.

Four preparatory operations were performed: operation 1, end-to-side anastomosis of the main PA to the ascending aorta at 8 months old (Melbourne shunt); operation 2, unifocalization of the right lower lobe MAPCA, modified Blalock-Taussig (B-T) shunt using a 5-mm polytetrafluoroethylene (PTFE) graft at 15 months

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