BRIEF COMMUNICATIONS

MEDIAN STERNOTOMY FOR PROLONGED RESUSCITATION IN NEONATAL TETRALOGY OF FALLOT WITH ABSENT PULMONARY VALVE

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Absent pulmonary valve syndrome is a subset of tetralogy of Fallot, with a reported prevalence of 2.6% to 6.0% among patients with tetralogy of Fallot.¹⁻³ This pathologic and clinical status is denoted by a vestigial pulmonary valve with pulmonary insufficiency; the pulmonary valve annulus frequently is moderately hypoplastic. Severe tracheobronchial compression may result from massive aneurysmal dilatation of the main pulmonary artery and its first- and second-order branches and from abnormal branching of segmental arteries.²⁻¹⁵ The degree of respiratory compromise and heart failure associated with left-to-right shunting are the chief determinants of the clinical presentation. Neonates at the severe end of the spectrum have an extremely high mortality rate despite aggressive attempts at medical and surgical management. In this report, we describe an alternative management option for this subset of absent pulmonary valve syndrome.

A 2.9 kg, 4-day-old male neonate with profound cyanosis and respiratory acidosis was found to have tetralogy of Fallot with absent pulmonary valve syndrome. Limited transthoracic echocardiographic examination demonstrated a massive dilatation of the main pulmonary artery (diameter 17 mm) and both branch pulmonary arteries (diameter 16 mm). Adequate ventilation could only be achieved with the patient in the prone position. Operative intervention was considered the only chance for survival of the patient.

The patient was repositioned from the prone to the supine position only shortly before performance of the median sternotomy. Sternotomy resulted in an immediate improvement in the mechanical gas exchange, as documented by a decrease in arterial carbon dioxide tension and an increase in arterial oxygen tension (Table I). Because of the gravity and the duration of the preoperatively existing metabolic derangement and a persistent moderate acidosis, however, it was decided to leave the sternotomy open without definitive repair to allow a prolonged period of resuscitation in the intensive care unit. After removal of the sternal retractor, there was a slight increase in the arterial carbon dioxide tension (Table I). It was therefore decided to leave the sternal retractor in place and to cover the sternotomy wound with Steri-Drape film (3M Company, St. Paul, Minn.). This

maneuver resulted in a stable hemodynamic situation and absence of arterial carbon dioxide retention in the supine position. This favorable clinical condition allowed for a detailed transesophageal echocardiographic examination. After 24 hours, a complete repair of the lesion, consisting of ventricular septal defect closure, aneurysmorrhaphy of the anterior and posterior walls of the dilated central pulmonary arteries, resection of the main pulmonary artery and vestigial pulmonary valve, and implantation of a valved pulmonary allograft to reconstruct the right ventricular outflow tract, was performed. The length of the allograft was kept short to lift the branch pulmonary arteries from the tracheobronchial structures.

The postoperative course was relatively uneventful. The patient was successfully weaned from the ventilator on the fifth postoperative day. There was constant subsequent improvement in respiratory status. At 6-month follow-up, the patient is doing well; another echocardiogram showed the right ventricular outflow tract and central pulmonary arteries to be of normal caliber.

Discussion. Despite early surgical correction of tetralogy of Fallot with absent pulmonary valve syndrome, mortality is still considerable. This high mortality rate is related mainly to the extremely poor clinical condition of these neonates and infants as a result of tracheobronchial compression and heart failure.^{1, 4, 5, 7-13} Nursing of the baby in a prone, head-up position is recommended as soon as the condition is recognized; this position allows the aneurysmally enlarged central pulmonary arteries to fall forward and away from the trachea and bronchi, particularly the right bronchus.^{9, 14, 15} Even if the patient responds well to this maneuver, we believe that operation should not be deferred. The only rational treatment for symptomatic absent pulmonary valve syndrome consists of correction of the anatomic substrate for the tracheobronchial compression and the intracardiac defect.

In the absence of severe respiratory compromise and secondary metabolic derangement, consideration should be given to early primary repair, including closure of the malalignment ventricular septal defect; reduction plasty of both the anterior and posterior walls of the main, left, and right pulmonary arteries; and valved allograft reconstruction of the right ventricular outflow tract. In the presence of severe respiratory compromise associated with acidosis and metabolic derangement, performance of a median sternotomy usually results in immediate improvement in gas exchange as a result of mechanical decompression of the tracheobronchial tree.¹⁴ Depending on the subsequent trend in clinical improvement in terms of gas exchange and resolution of acidosis, as observed in the operating

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Table I. Respiratory and metabolic status data

Variable	Intraoperative					
	Preoperative		Before sternotomy		After sternotomy	
	Supine	Prone	Prone	Supine	Without retractor (20 min)	With retractor (30 min)
PCO ₂ (mm Hg)	68	39	40	72	42	34
PO ₂ (mm Hg)	25	60	69	65	340	375
pH	7.21	7.33	7.34	7.25	7.32	7.38
Fio ₂	0.4	0.4	0.4	1.0	1.0	1.0
Ventilation	SIMV	SIMV	SIMV	CMV	CMV	CMV

PCO₂, Partial pressure of carbon dioxide in arterial blood; PO₂, partial pressure of oxygen in arterial blood; Fio₂, fraction of inspired oxygen; SIMV, synchronized intermittent mandatory ventilation; CMV, continuous mandatory ventilation.

room, the surgeon must judge whether the clinical status of the baby allows primary repair of the underlying defect at that time or whether a prolonged period of resuscitation in the intensive care unit is likely to be necessary. If, as in the case of our patient, prolonged resuscitation is selected as the management option, the sternotomy wound is temporarily closed with a prosthetic patch such as a silicone rubber patch reinforced with Dacron polyester fabric mesh or a polytetrafluoroethylene patch. Alternatively, in the presence of severe tracheobronchial compression that is incompletely relieved by median sternotomy, the sternal retractor can be left in place to aid in continuous decompression and the wound can be sealed with Steri-Drape plastic film.¹⁶ Such staged management may reduce the risk of the subsequent operation in these critically ill patients, and it allows further diagnostic studies if needed. Complete repair of the underlying anomaly can then be undertaken after a period of 24 to 72 hours.

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