CASE REPORT

Postoperative Prone Position Management of Tetralogy of Fallot with Absent Pulmonary Valve Syndrome

Tetralogy of Fallot with absent pulmonary valve syndrome is commonly associated with respiratory failure both before and after surgery. This report describes our experience using prone positioning with bilateral pillows to avoid compression of the anterior chest wall after surgery. In the case here, the patient's respiratory distress was improved by this positioning. Prone position and avoiding anterior chest compression has an effect on severe respiratory distress of tetralogy of Fallot with absent pulmonary valve syndrome. (Jpn J Thorac Cardiovasc Surg 2005; 150–153)

Key words: ventilation, prone position, tetralogy of Fallot, absent pulmonary valve syndrome, carbon dioxide retension

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Tetralogy of Fallot (TOF) with absent pulmonary valve syndrome (APVS) is commonly associated with tracheobronchial compression caused by aneurysmal dilatation of the main pulmonary artery (PA) and its branches.[1,2] The ventilatory status of these patients can be unstable both before[3-5] and after surgery. This report describes our experience using prone positioning to avoid compression of the anterior chest wall after surgery.

Case

A 2.3-kg female neonate was referred on the first day of life for evaluation and treatment for respiratory failure and esophageal atresia. At birth, endotracheal intubation and mechanical ventilation were necessary. Further evaluation revealed TOF with APVS, esophageal atresia and tracheo-esophageal fistula, duodenal atresia, hydrocephalus, aplastic kidney, and laryngomalacia. Respiratory failure occurred by bronchial stenosis due to dilated PA of TOF with APVS. She underwent surgical repair of the esophageal atresia and tracheo-esophageal fistula, and duodenal atresia in the neonatal period. Postoperatively, although mild bronchial compression existed, her ventilatory status was stable without mechanical ventilation and at ten weeks of age, she was discharged home.

At six months of age, she was admitted for respiratory failure; endotracheal intubation and mechanical ventilation were necessary. Her weight on admission was 4.7 kg. A chest roentgenogram showed a cardiothoracic ratio of 45% and bilateral pulmonary emphysema. Pressure data were as follows: left ventricle 87/11 mmHg, right ventricle 85/7 mmHg, and main PA 28/9 mmHg. Qp/Qs was 1.7. Chest computed tomography showed right and left bronchial stenosis and a compressed and dilated PA (Fig. 1). We assessed the main cause of respiratory failure as progressive severe bronchial compression and pulmonary emphysema. She underwent cardiac surgery, which included closure of the ventricular septal defect and aneurysmorraphy of the massively dilated main PAs, which involved resection of the anterior wall with plication of the posterior wall of both PA branches.

Postoperatively, she was stable with low dose catecholamine. She had no carbon dioxide (CO_2)

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Received for publication May 26, 2004.

Accepted for publication November 15, 2004.

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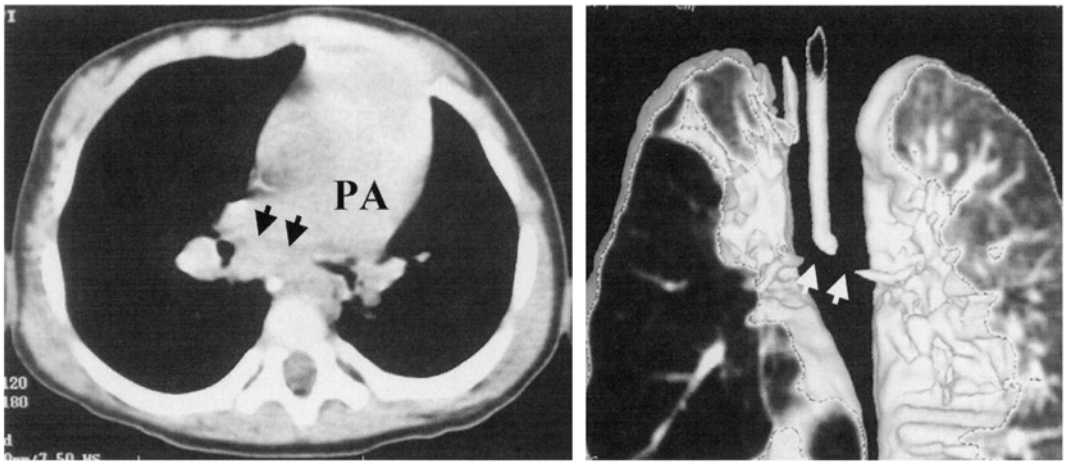

Fig. 1. Preoperative chest computed tomography shows right and left bronchial stenosis (arrows).
PA, Pulmonary artery.

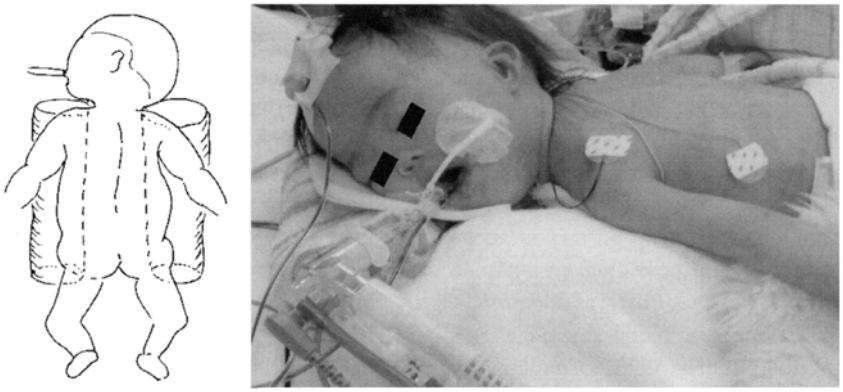

Fig. 2. Two pillows to avoid anterior chest compression.

retention in the supine position during the first 24 hours. However two days later, she developed severe CO_2 retention and did not respond to either bronchodilators or deep sedation. Adequate ventilation could only be achieved with the infant in the prone position. Blood gas analysis showed normal CO_2 levels. Twelve hours later, however, she developed right heart failure, with elevated central venous pressure and decreased blood pressure and urine output. Two cushion-like pillows were put between the shoulder and thigh on each side to prevent anterior chest compression (Fig. 2). Subsequently, her respiratory and hemodynamic status became stable. The Table I shows respiratory and hemodynamic status data, and the mechanical ventilation set up varying with positioning. Four days later her position was changed back to supine again. Attempts to wean the patient from the ventilator were unsuccessful because of laryngomalacia and bronchial stenosis. Due to the multiple anomalies, ventilatory support was necessary for a long time. She was eventually extubated on day 90 postoperatively. Eventually, the patient was discharged and did very well. At the six-month follow-up, she remains well and free of respiratory symptoms.

Discussion

APVS is usually associated with TOF, which has a clinical course of severe respiratory distress caused by compression of the bronchial tree by the markedly dilated PA. Improvement of the respiratory distress by positioning infants with symptomatic APVS in the prone position has been noted. This positioning decreases the compression of the bronchi caused by dilated PA, producing the improvement. Therefore, such patients

Table I. *Respiratory and hemodynamic status data varying with positioning*

	Arrival ICU supine	*CO_2 retention supine*	*Immediate after prone*	*Prone 6 hours later*	*Prone without chest compression*
PCO_2 (mmHg)	46	66	34	45	39
PO_2 (mmHg)	199	88	141	332	286
pH	7.38	7.29	7.47	7.38	7.41
CVP (mmHg)	9	10	7	12	7
Systemic BP (mmHg)	80	85	82	55	108
Urine output (ml/hr)	14	3	10	0	20
FiO_2	0.7	0.4	0.3	0.9	0.8
PIP (cm H_2O)	20	22	20	21	21
PEEP (cm H_2O)	0	0	0	1	1
RR (/min)	20	25	25	25	25

ICU, Intensive care unit; *CVP,* central venous pressure; *BP,* blood pressure; *PIP,* peak inspiratory pressure; *PEEP,* positive end-expiratory pressure; *RR,* respiratory rate.

need to be placed in the prone position, even postoperatively, although postoperative patients may not tolerate the prone position because of anterior chest and right ventricular outflow compression. When it is necessary for a postoperative patient to be in the prone position, the possibility of heart failure due to anterior chest compression need to be considered.

Mechanical ventilation was set up on arrival at the intensive care unit (ICU) with the patient in a supine position. Peak inspiratory pressure (PIP) was 20 cm H_2O, respiratory rate (RR) 20/min and blood data analysis at that time showed PCO_2 46 mmHg, PO_2 199 mmHg. To increase inspiratory volume, we changed the set up to PIP 22 cm H_2O, RR 25/min, because of progressive CO_2 retention in the supine position. Despite increased PIP and RR, PCO_2 was 66 mmHg and PO_2 88 mmHg indicating lower ventilation. During this period, chest X-ray showed bilateral emphysema; the same finding existing before the radical operation. There were no atelectasis, pleural effusion, pneumothorax, or pneumonia. We assessed the main causes of progressive CO_2 retension to be tracheal and bronchial stenosis due to: 1) the compression of the trachea and main bronchus due to gravitational PA weight, and 2) tracheal and main bronchial collapse due to their own gravitational weight. We think that postoperative tracheal and bronchial collapse occurred by edematous change due to capillary leakage during cardiopulmonary bypass, and inflammation and edema caused by stimulation of the intratracheal suction tube.

In a radical operation for TOF with APVS, PA placation creates vacant spaces preoperatively occupied by the huge PA in the anterior of the trachea and bronchus: thus, prone positioning would be more effective in that the trachea, bronchus, and PA would be attracted to the vacant anterior spaces. Although postoperative tracheal and bronchial stenosis by PA compression can occurr in other diseases with a large PA due to preoperative high pulmonary flow, the prone position would be less effective to perform the operation without PA placation. Pillows to prevent anterior chest compression could increase the vacant spaces in the anterior of the trachea and bronchus after PA placation for TOF with APVS. We think that patients with respiratory management difficulties in a supine position due to severe tracheal and bronchial stenosis by preoperative PA compression after PA placation for TOF with APVS should be placed in a prone position using pillows.

In conclusion, we undertook postoperative prone-position management of TOF with APVS, with the result that CO_2 retention improved. In the case presented here, pillows placed bilaterally to avoid anterior chest compression, instigated a dramatic improvement in respiratory and hemodynamic status. Prone position and avoiding anterior chest compression can have an effect on severe respiratory distress in TOF with APVS.

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