Case Report

Preoperative Respiratory Physiotherapy for a Patient with Severe Respiratory Dysfunction and Annuloaortic Ectasia

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A 23-year-old man with Marfan syndrome, who had undergone surgery for pectus excavatum and scoliosis and who had severe respiratory dysfunction, was referred for surgical repair of annuloaortic ectasia. The preoperative pulmonary function test revealed severe obstructive and restrictive respiratory dysfunction, with forced expiratory volume in one second of 650 ml and vital capacity of 1,220 ml. These parameters improved after 4 months respiratory physiotherapy. A modified Bentall's procedure was performed after respiratory physiotherapy. A tracheostomy made on the 7th postoperative day (POD) appeared to improve respiratory condition and he was weaned off mechanical ventilation on the 14th POD. The lower limits of pulmonary function for open heart surgery have not been established clearly; however, our case will help elucidate these limits of respiratory function for open heart surgery. Preoperative respiratory physiotherapy improved parameters of pulmonary function test and may decrease the morbidity of postoperative pulmonary complications in a patient with severe respiratory dysfunction. (Ann Thorac Cardiovasc Surg 2003; 9: 266–9)

Key words: Marfan syndrome, annuloaortic ectasia, Bentall's procedure, respiratory dysfunction, pectus excavatum, scoliosis

Introduction

Marfan syndrome is an autosomal dominant inheritable disorder that affects connective tissue. It is characterized by abnormalities in the cardiovascular system, skeleton, and eyes.[1)] Many patients with Marfan syndrome referred for cardiovascular surgery have some degree of chest deformity such as funnel chest, which is known to affect pulmonary function.[2)] We describe a patient with Marfan syndrome who had severe respiratory dysfunction and discuss efficacy of preoperative physiotherapy and lower limits of pulmonary function in patients undergoing open heart surgery.

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Case Report

A 23-year-old man with Marfan syndrome was admitted to our hospital for surgical repair of annuloaortic ectasia. His family history indicated the death of a brother from rupture of an abdominal aortic aneurysm. His previous treatment included 1) surgery for Hirschsprung disease at the age of 1, 2) a turnover procedure for pectus excavatum at 11, which recurred afterward, and 3) posterior spinal fusion for scoliosis at 17. The patient developed dyspnea on exertion for 1 year prior to the current admission. He had arachnodactyly and his constitution was typical of Marfan syndrome (height, 185 cm; weight, 45.2 kg). He had subluxation lentis and myopia. A diastolic murmur was heard on the third left intercostal space.

An electrocardiogram showed normal sinus rhythm with incomplete right bundle branch block. A chest radiograph disclosed cardiomegaly (cardiothoracic ratio, 65%), widening of the mediastinum, funnel chest, and scoliosis (Fig. 1). Echocardiography revealed dilatation of the ascending aorta at the root (41 mm) with aortic regurgitation (grade II) and left ventricular dilatation. The

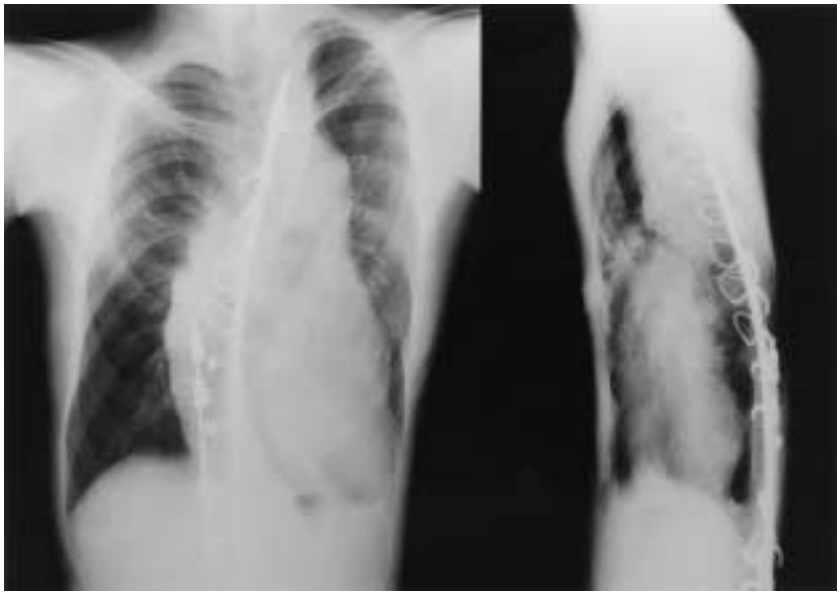

Fig. 1. Remarkable chest deformity with mild cardiomegaly was shown on the chest X-ray film.

pH of the arterial blood was 7.358, the partial pressure of oxygen (PO_2); 61.1 torr, carbon dioxide (PCO_2); 55.5 torr, bicarbonate; 30.5 mmol/L, base excess; 4.0 mmol/L with the patient breathing room air. A computed tomographic (CT) scan showed dilatation of the ascending aorta from the root with the maximum diameter of 7 cm (Fig. 2). The heart was shifted into the left chest cavity. Aortography was also performed with the finding of dilatation at the sinuses of Valsalva and the ascending aorta with aortic regurgitation. The aortic regurgitation was graded as III according to Seller's classification.

A preoperative pulmonary function test revealed obstructive and severe restrictive respiratory dysfunction. Preoperative respiratory physiotherapy, including incentive spirometry, diaphragm inhibition, and pursed-lip breathing was administered and parameters of respiratory function improved after 4 months physiotherapy (Table 1). Especially, maximal voluntary volume increased greatly; from 22.4 to 38.2 L/min. After 4 months of respiratory physiotherapy, the operation was performed with a median sternotomy. It was recognized on the CT scan that the superior vena cava existed just behind the sternum, so the sternum had to be carefully dissected from the tissue under it. A modified Bentall's procedure was carried out with a composite graft composed of a 23-mm CarboMedics mechanical valve (CarboMedics Inc., Austin, TX, USA) and a 26-mm Hemashield vascular graft (Meadox Medicals Inc., Oakland, NJ, USA). An 8-mm Hemashield vascular graft was used as a coronary conduit. We did not perform surgical treatment for recurrent pectus excavatum. The extracorporeal circulation time was

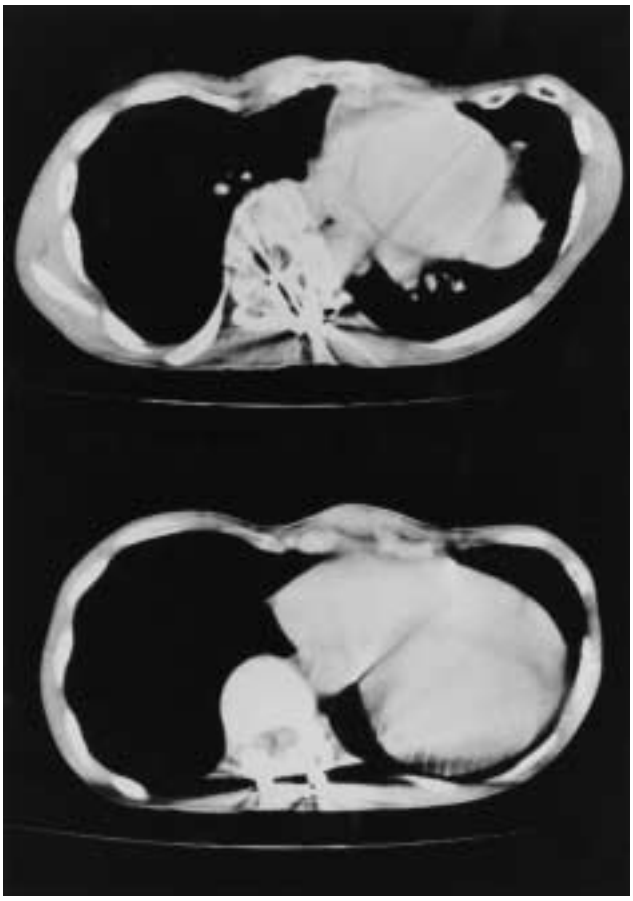

Fig. 2. Computed tomographic scan demonstrated dilatation of the ascending aorta from the root. The maximum diameter of the aorta was 7 cm. The heart was shifted into the left chest cavity.

Table 1. Pulmonary function test before and after respiratory physiotherapy and after surgery

Duration of respiratory physiotherapy	VC (Liter)	%VC (%)	$FEV_{1.0}$ (Liter)	$FEV_{1.0\%}$ (%)	%MVV (%)	$FEF_{25-75\%}$ (%)
Before	1.22	26.2	0.65	56.5	20.0	9.0
1 month	1.20	25.8	0.72	56.3	27.1	8.1
2 months	1.31	28.1	0.61	47.7	28.3	6.5
3 months	1.36	29.2	0.80	58.8	34.1	10.4
4 months	1.57	33.7	0.73	52.1	29.8	7.4
Postop	1.40	30.0	0.84	66.1	34.1	9.2

VC, vital capacity; %VC, percent predicted normal value of VC; $FEV_{1.0}$, forced expiratory volume in one second; $FEV_{1.0\%}$, $FEV_{1.0}$/forced vital capacity; MVV, maximal voluntary ventilation; $FEF_{25-75\%}$, forced expiratory flow between 25% and 75%; postop, examination one month after the operation.

3 hours and 5 minutes. The aortic cross-clamping time was 2 hours and 34 minutes. Just before and after the cardiopulmonary bypass (CPB), PO_2 and PCO_2 were measured under an inspiratory oxygen friction of 1.0. PO_2 and PCO_2 were 414 and 34 torr preoperatively and 429 and 48 torr postoperatively. Deterioration of pulmonary oxygenation capacity due to CPB was not recognized just after the termination of CPB.

The endotracheal tube was removed on the 1st postoperative day (POD), but reintubation was required because of hypercapnea and difficulty in sputum discharge. After a tracheostomy was performed on the 7th POD, his respiratory condition ameliorated, and he was weaned off the mechanical ventilation on the 14th POD.

Parameters of the pulmonary function test 1 month after the operation had not deteriorated (Table 1), and his respiratory status improved from grade 4 to grade 3 according to the Hough-Jones classification.

Discussion

The incidences of pectus excavatum and Marfan syndrome are 8 per 1,000 live births and 4 to 6 per 100,000 live births, respectively. However, pectus excavatum is often manifested in Marfan syndrome. It is reported that 68% of patients with Marfan syndrome have pectus excavatum.[1)]

Cahill et al. stated that patients with Marfan syndrome have impaired cardiorespiratory function.[2)] Therefore, surgical repair for pectus excavatum is performed to intend to improve cardiac and pulmonary functions as well as for cosmetic reasons. However, Arn et al. emphasized that surgical repair of pectus excavatum in patients with Marfan syndrome should be delayed if possible until skeletal maturity is nearly complete. Otherwise, the pectus deformity recurrs frequently (40%).[3)] The patient in this case had surgical repair for pectus excavatum at age 11, and after 12 years severe pectus deformity recurred. Scoliosis is also related to pulmonary dysfunction.[4)] These two concomitant skeletal malformations of Marfan syndrome result in severe respiratory dysfunction, especially restrictive respiratory dysfunction.

Surgical strategy must be considered for annuloaortic ectasia with concomitant pectus excavatum. One option is a staged repair to avoid prolonged operative time and excessive skeletal bleeding under the use of full anticoagulation during CPB. Recently, however, successful simultaneous repairs of pectus excavatum and cardiovascular abnormalities have been reported.[5)] This means excessive bleeding can be prevented by recent pharmacological improvement, such as aprotinin and gelatin resorcin formal (GRF) glue, as well as by refinement of surgical procedure. In this case, even though aprotinin was used, only surgical repair of annuloaortic ectasia was performed because we thought that surgical repair of the recurrent pectus excavatum would take more operative time and have excessive bleeding and require large amounts of transfusion, which would make the postoperative pulmonary function worse and make it difficult to wean off from a mechanical respiratory support.

The lower limits of pulmonary function for open heart surgery have not been established. A prospective study demonstrated maximal voluntary ventilation (MVV) and forced expiratory flow in 25%-75% ($FEF_{25-75\%}$) of less than 50% of the predicted normal mean and forced vital capacity (FVC) of less than 75% of the anticipated normal value are indications of a high probability of serious postoperative pulmonary complications.[6)] The patient was categorized as being at high risk for serious postoperative pulmonary complications according to the criteria mentioned above and actually had prolonged mechanical ventilation because of respiratory failure. However, it is

noteworthy that, even though the patient had postoperative respiratory failure, we were able to wean him off the mechanical ventilator without incidence of pneumonia, and he now lives without any respiratory support. After 4 months respiratory physiotherapy %MVV improved 1.5 times. We believe that the preoperative respiratory physiotherapy itself contributed to the successful postoperative weaning off from mechanical respiratory support.

Open heart surgery in patients with more impaired respiratory function most likely will become more common because of an increasing aged population. Thus, it is more important than ever to determine the lower limits of pulmonary function in open heart surgery. Until now many investigations have been attempted, but clear criteria, including limit of parameters in pulmonary function tests, have not been defined.

It is well known that CPB causes lung injury, and recent studies of cytokines as well as complements are helping elucidate the mechanism of lung injury that accompanies CPB. To prevent lung injury during open heart surgery, shortening of extracorporeal circulation time and a decreased amount of transfusion are believed to be crucial. In addition, pulmonary congestion is caused by CPB, and therefore, urination is an essential component of care after open heart surgery in patients with respiratory dysfunction. These facts mean that other parameters of pulmonary function would also be related to the morbidity of pulmonary complications after open heart surgery. Some pharmacological treatments such as an elastase inhibitor have been developed to prevent lung injury during CPB.[7] The possibility of pharmacological prevention of lung injury is likely to widen the indication for open heart surgery for patients with more impaired respiratory function.

It is debatable whether postoperative tracheostomy was required or not. It is reported that a tracheostomy after a median sternotomy increases the incidence of mediastinitis. In our case, the tracheostomy was performed a week after the median sternotomy, but fortunately mediastinitis was avoided because of enough distance between the median sternal wound and the wound at the site of the tracheostomy. A tracheostomy for patients with respiratory failure is believed to be better than nasotracheal intubation with respect to sputum discharge.[8] We thought tracheostomy was necessary to the patient because difficulty in sputum discharge resulted in respiratory failure. Otherwise, he might have higher risk of pulmonary complications. It is generally accepted that preoperative pulmonary preparation, including nebulizer and respiratory physiotherapy, decreases postoperative morbidity. However, physiotherapy for patients with heart failure or aortic aneurysm seems to be dangerous. We performed the respiratory physiotherapy with great caution. The program of the physiotherapy was well arranged with discussion between surgeons and physiotherapists. The patient did not have any trouble during the physiotherapy except for chest pain a few times. When he felt discomfort or chest pain, the physiotherapy was ceased and physical assessments including electrocardiography were done. The progression of aortic aneurysm and aortic valve regurgitation was also evaluated during the physiotherapy. The efficacy of preoperative respiratory physiotherapy has not been fully evaluated and further studies are required to know to what extent respiratory physiotherapy can be done safely for patients with heart diseases including heart failure or aortic aneurysm and to what extent respiratory physiotherapy should do to decrease the morbidity of pulmonary complications.

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