



THE ANNALS OF THORACIC SURGERY



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Ann Thorac Surg 2003;75:1304-1306

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not to release emboli that may affect nearby vascular structures (carotid, ophthalmic, and so forth) [10]. Therefore it is fundamental to choose a good material for embolization. One must avoid liquid solutions and particles less than 250 microns in size caused by the major risk of spread through neighboring vascular structures and causing medullary lesion or tissular necrosis [4], or both. The use of coils or occlusive balloons is not recommended either, as this only causes proximal occlusion of the vessel with the possible formation of collaterals that re-channel the distal bed [4].

In the cases described of embolizations for hemoptysis, it is brought under control in more than 90% of patients [7–8], with its major limitation being the high rate of early recurrence (14% to 20%) [6, 9, 11]. This is attributed to an incomplete embolization or re-channelling of the treated vessels, which means an exhaustive search must be made for all vessels potentially responsible for the bleeding, as in our case [11]. The late recurrence rate is also high (18% to 28%) [9, 11] and generally represents progression of the underlying pulmonary disease. The embolization was final in our case, because all vascularization of the hemangioma was embolized, thus achieving total remission of the same. If there is no base disease other than the hemangioma (eg, tuberculosis), it can be considered cured.

In conclusion, tracheal capillary hemangioma may produce a massive hemoptysis that can be controlled efficiently and definitively with embolization by interventional radiology.

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Hemopneumothorax Associated With Marfan Syndrome and Congenital Afibrinogenemia

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In patients with afibrinogenemia who require operation, prophylaxis against bleeding is important. We report the case of a 14-year-old boy with Marfan syndrome and congenital afibrinogenemia in whom hemopneumothorax developed. Video-assisted thoracoscopic surgery was performed successfully under intravenous administration of fibrinogen and with careful monitoring of plasma fibrinogen level.

(Ann Thorac Surg 2003;75:1304–6)

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Congenital afibrinogenemia is a rare, inherited coagulopathy with clinical manifestations that range from minor to severe bleeding because of the complete absence of fibrinogen. Marfan syndrome is a heritable connective tissue disorder with manifestations in the ocular, skeletal, and cardiovascular systems. Marfan syndrome is often associated with pneumothorax. We report the first case of hemopneumothorax, recurrent bilateral pneumothorax with Marfan syndrome, and congenital afibrinogenemia. Video-assisted thoracoscopic bullectomy was successfully performed by intravenous administration of heat- and solvent detergent-treated fibrinogen concentrates.

A 14-year-old boy was admitted to our hospital complaining of chest pain and dyspnea. He was known to have congenital afibrinogenemia, diagnosed 4 days after birth following umbilical bleeding. The bleeding episodes included submandibular bleeding and subdural hematoma at the age of 2 years. Marfan syndrome was diagnosed at age 12 on the basis of physical stigmata and cardiovascular abnormalities. He had been treated with 1 g of fibrinogen intravenously every 2 weeks since age 6 years and treated for chronic hepatitis due to hepatitis C virus infection since age 11 years.

A chest roentgenogram and computed tomographic scan on admission showed collapse of the left lung and fluid collection (Fig 1). He was found to have hemopneumothorax by chest radiography and was treated with pleural drainage. About 1,100 mL of fresh blood was drained. The plasma fibrinogen level was 15 mg/dL. He

Accepted for publication Oct 8, 2002.

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A



B

Fig 1. Chest roentgenogram (A) and computed tomographic scan (B) on admission showing collapse of the left lung and pleural effusion.

was treated with fibrinogen, which resulted in cessation of bleeding. The chest drainage tube was removed within a few days. However, 1 month later, recurrent left pneumothorax developed. We decided to perform thoracoscopic bullectomy. Five grams of fibrinogen was infused during 3 preoperative days, which increased the plasma fibrinogen level to 183 mg/dL. Then he underwent thoracoscopic bullectomy. We found a bulla measuring 2.0 cm in diameter at the apex of the left upper lobe. The bulla was resected by stapling device. Two grams of fibrinogen were infused daily until postoperative day

(POD) 3, and 1 g of fibrinogen was infused on POD 4 and POD 5. The fibrinogen level was 154 mg/dL on POD 3 (Fig 2). His postoperative course was uneventful, and there was no postoperative bleeding. The chest tube was removed on POD 2, and the patient was discharged on POD 7. However, left pneumothorax recurred at 16 years of age and right pneumothorax developed 2 years later. He was treated successfully by bilateral thoracoscopic bullectomy using the same perioperative management protocol. We have followed up this patient for 4 years after the last operation, and he remains free from any episode of pneumothorax or hemopneumothorax.

Comment

Congenital afibrinogenemia is a rare inherited disorder with an estimated incidence of about 1 to 2 per million. Clinical manifestations of this disorder range from minimal bleeding to fatal bleeding [1]. Almost all bleeding episodes are treated conservatively with intravenous administration of fibrinogen. However, some cases of spontaneous splenic rupture and surgical treatment associated with afibrinogenemia have been reported [2, 3]. Congenital afibrinogenemia is caused by mutation of fibrinogen alpha and beta chain gene [6].

Marfan syndrome is an autosomal dominant inherited disorder of connective tissue with variable clinical manifestations in the cardiovascular, ocular, musculoskeletal systems and is often complicated by pneumothorax [4]. Our case represents rare and complicated condition. Recently the molecular pathogenesis of Marfan syndrome was recognized resulting from mutation in the fibrillin-1 (FBN1) gene [5]. The genetic relationship between these two inherited disorders is not known.

In patients with afibrinogenemia, prophylaxis of bleeding is important if they need an operation. However, fibrinogen therapy is associated with the risk of viral infections, such as hepatitis viruses and human immunodeficiency virus. In addition, allergic reactions and anti-fibrinogen antibody have been reported [3]. We commonly use heat- and solvent detergent-treated fibrinogen, but the risk of viral

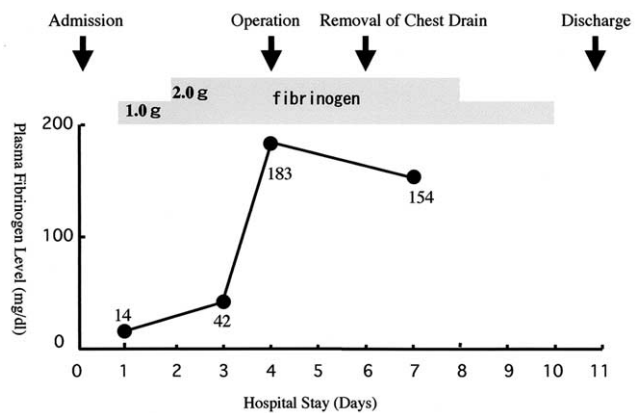


Fig 2. Serial changes in plasma fibrinogen concentration during hospitalization.

infection remains. Unfortunately, our patient developed chronic hepatitis due to hepatitis C virus. It is possible that he had been infected by blood products before the introduction of the viral screening system.

Fibrinogen must be administered with continuous monitoring of plasma fibrinogen level during the perioperative period. For prophylaxis of perioperative bleeding, the fibrinogen level must be over 100 mg/dL [1-3]. In our case, 7 g of fibrinogen was administered preoperatively and 8 g postoperatively. Such treatment resulted in maintenance of plasma fibrinogen levels above 150 mg/dL during the perioperative period, and the operation was performed safely.

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Cardiac Fibroma in an Infant: Single Ventricle Palliation as a Bridge to Heart Transplantation

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A prenatal echocardiogram revealed a large right ventricular mass. Following birth, there was obstruction to pulmonary blood flow and cyanosis. The tumor's size and location prevented resection. The patient underwent "single ventricle palliation," including placement of a systemic-to-pulmonary shunt as a newborn. This palliation served as a successful bridge to heart transplantation at 7 months of age. Pathologic examination revealed cardiac fibroma.

(*Ann Thorac Surg* 2003;75:1306-8)

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Accepted for publication Oct 14, 2002.

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Cardiac fibroma is the second most common cardiac tumor of childhood (after rhabdomyoma). Although benign, fibromas may cause chest pain, arrhythmias, or, if large enough, obstruction to blood flow with congestive heart failure or cyanosis [1, 2]. Symptomatic fibromas have been successfully treated by resection, either partial or complete, or by heart transplantation [1, 3-8]. We present the case of a neonate with an unresectable right ventricular fibroma and cyanosis. He underwent "single ventricle palliation," which served as a successful bridge to heart transplantation.

Fetal echocardiography at 33 weeks' gestation revealed a large right ventricular mass and a pericardial effusion. Following delivery at 37 weeks, physical examination was notable for slight prominence of the right hemithorax, and a grade 2/6 systolic ejection murmur along the left sternal border. Chest roentgenogram illustrated a large cardiac silhouette extending well into the right chest and obscuring the right lung. Echocardiography revealed a mass occupying most of the right ventricle and atrium. There was right-to-left flow across a small, restrictive patent foramen ovale, and minimal antegrade flow across the right ventricular outflow tract. Pulmonary flow was provided by a patent ductus arteriosus. A large pericardial effusion was removed by pericardiocentesis and prostaglandin E-1 was administered to maintain ductal patency. Cardiac magnetic resonance imaging (MRI) verified a large mass centered in the right ventricle with compression of the right atrium and much of the right lung. Attempts to wean the prostaglandin were unsuccessful due to hypoxia.

As the tumor appeared unresectable, it was decided to carry out "single ventricle palliation," consisting of atrial septostomy and placement of a systemic-to-pulmonary artery shunt. The aim of this palliation was to allow weaning from prostaglandin E-1, and to serve as a possible bridge to heart transplantation. Balloon atrial septostomy was successfully performed in the cardiac catheterization lab. Angiography confirmed near obliteration of the right ventricular cavity and indicated that the right coronary artery was draped over the center of the mass. At 1 week old the patient was explored through a median sternotomy. A large, firm mass was contained within the pericardium, but had expanded to fill most of the mediastinum and right pleural space. The mass occupied the entire anterior surface of the right ventricle, displaced the left ventricle leftward, and prevented access to the right atrium, vena cavae and right pulmonary artery. A 4-mm ringed Gore-Tex shunt (W.L. Gore & Associates, Flagstaff, AZ) was placed from the base of the innominate artery to the main pulmonary artery and the ductus arteriosus was ligated. Biopsies of the mass characterized a spindle cell tumor with collagen deposition and no mitotic activity. Postoperatively the patient was successfully extubated with a systemic oxygen saturation of 87% on room air. As the natural history of his tumor was unclear, he was discharged and followed with serial echocardiograms.

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