



# Extrapleural pneumonectomy in children

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## Abstract

**Background/Purpose:** Extrapleural pneumonectomy (EPP) is an en bloc resection of the pleura, lung, diaphragm, and pericardium. EPP in the pediatric population has not been reported in the literature. We report our experience of using EPP in children to treat a variety of malignancies that involve the pleural surface.

**Methods:** We performed a retrospective review of all children treated through EPP at our institution. Data were obtained from patient charts and a pediatric database. All patients underwent en bloc resection of the pleura, lung, diaphragm, and pericardium and reconstruction.

**Results:** Four patients underwent EPP from 2000 to 2004 for inflammatory myofibroblastic tumor, spindle cell sarcoma, metastatic neuroblastoma, and malignant mesothelioma. The patients' ages were from 6 to 11 years. The patient with metastatic neuroblastoma had recurrent disease in the left chest 1 year after EPP and died of sepsis; the patient with mesothelioma died of disease extension into the abdomen 1.5 years later. The other 2 patients are free of disease.

**Conclusions:** EPP can be performed with curative intent in selected pediatric patients with pleural-based malignancies. EPP should be included in the armamentarium of the pediatric thoracic surgeon and evaluated in larger studies.

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Pneumonectomy in children is performed for both neoplastic and infectious diseases. Although infectious etiology is the most common indication for pneumonectomy in developing countries [1-3], congenital anomalies have replaced tuberculosis and bronchiectasis as the more common reason for pneumonectomy in the pediatric population of developed nations. In addition, primary or metastatic pulmonary tumors may also require pneumonectomy. Extrapleural pneumonectomy (EPP) is useful for pleural-based lesions localized to the hemithorax. EPP has been studied extensively in adults, especially for the

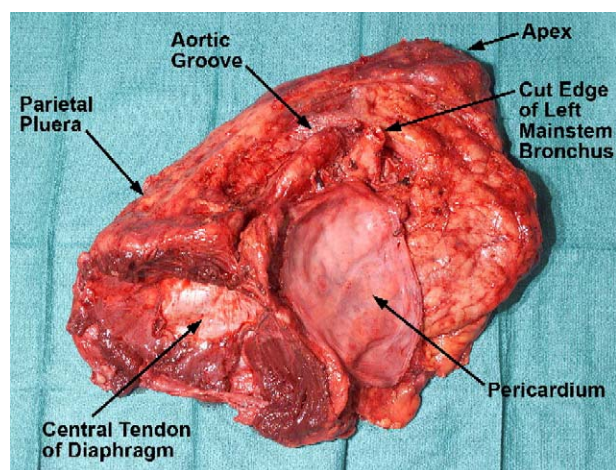
treatment of malignant pleural mesothelioma, but EPP in children has not been reported. We described a series of 4 patients undergoing EPP for malignant disease.

## 1. Materials and methods

### 1.1. Data acquisition

A pediatric surgery database was reviewed; all children undergoing EPP from June 2000 to October 2004 were included and, data were collected with Institutional Review Board approval. EPP was defined as an en bloc resection of the pleura, lung, diaphragm, and pericardium (Fig. 1) [4,5]. Patients with only intrapleural or intrapericardial pneumonectomies were excluded.

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**Fig. 1** Photograph of resected en bloc specimen demonstrating intact pleural envelope consisting of pleura, pericardium, diaphragm, and lung.

## 1.2. Surgical technique

Airway management included the use of bronchial blockers in 3 patients and a double lumen endotracheal tube in the oldest patient. EPP was performed via an extended posterolateral thoracotomy incision, and the best exposure was gained when the anterior portion of the incision is carried to the costal margin [6]. The chest is entered through the fifth interspace, and the extrapleural plane, which lies between the endothoracic fascia and the outer surface of the parietal pleura, can be developed by a combination of both blunt and sharp dissection. Although resection of the sixth rib is routinely performed in adults, it is not necessary in the pediatric patient because of a very elastic chest wall.

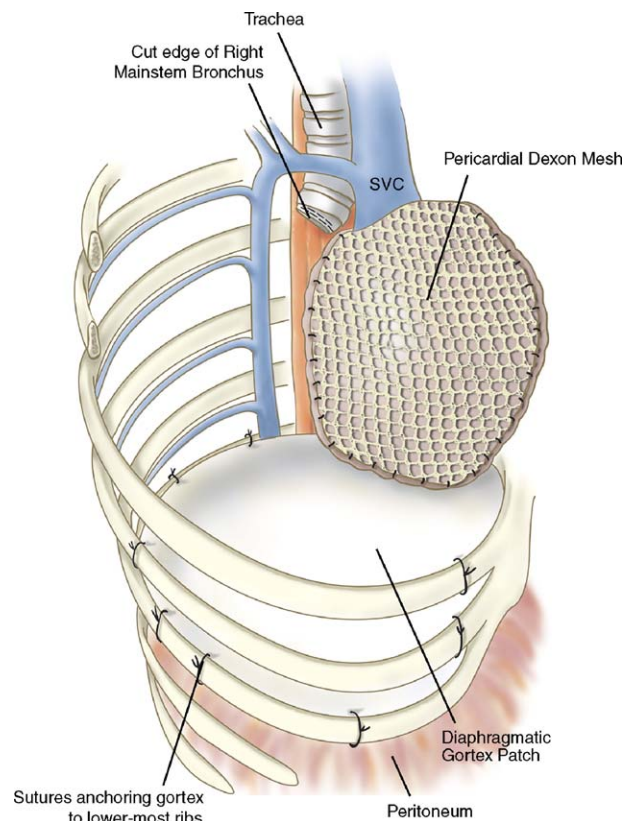
Once enough parietal pleura have been mobilized, chest retractors are inserted to help with exposure. Because this is a very vascular plane, it requires continuous packing with lap pads and use of the argon beam electrocoagulator to minimize blood loss. The dissection is carried superiorly to the subclavian vessels. Care must be taken near the mammary vessels because these may be avulsed at their origin from the subclavian vessels. On the left side, care must be taken not to injure the esophagus or the aorta—especially the intercostal branches. On the right side, the superior vena cava may be adherent to the pleura. There may be a distinct plane of dissection between the anterior portion of the hilum and the pericardium in some cases. In others, this plane is obliterated, and an en bloc resection with the pericardium is required.

The lower hemithorax and diaphragm are then dissected. A ridge can be palpated at the junction of the tumor and the diaphragmatic muscle. Once the diaphragm is mobilized from the posterior costophrenic angle, it is rotated into the thoracotomy incision, thereby allowing exposure of the costophrenic sulcus and further dissection posteriorly. If involvement of the diaphragm is extensive, it should be

removed in its entirety. If involvement of the diaphragm is superficial, dissection may be carried through the diaphragmatic muscle using electrocautery. The peritoneum should be peeled away from the back of the diaphragm by means of a sponge stick or lap pad. Because tissue planes (peritoneum and diaphragm) become confluent at the level of the central tendon of the diaphragm, the peritoneum is often opened during this part of the dissection. It should be immediately reclosed with absorbable suture to prevent tumor implantation into the peritoneal cavity, especially in patients with mesothelioma.

Once the entire extrapleural lung and diaphragm are completely mobilized, the pericardial interface is assessed. Depending on the central extent of the tumor, the hilar vessels are transected intrapericardially or may be transected in the extrapericardial extrapleural plane. The specimen consisting of pleura, lung, and diaphragm, with or without pericardium, is removed en bloc.

The diaphragm is then reconstructed on the right side, and a double-layer Dexon mesh (US Surgical: Norwalk, Conn) is preferred because the liver helps to prevent herniation of intraabdominal contents. On the left side, 2-mm-thick Gore-Tex (W.L. Gore and Associates, Inc, Flagstaff, Ariz) is used because it is more durable at resisting herniation. The prosthesis is secured by placing sutures around the ribs laterally (Fig. 2). It is secured to the crus posteriorly and sewn to the edge of the pericardium



**Fig. 2** Pericardial and diaphragmatic reconstruction.

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