

Foregut Duplication Cysts in Children

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ABSTRACT

Background and Objectives: Duplications of the alimentary tract are rare anomalies. We report our experience with foregut duplication cysts including their clinical presentation, diagnostic modalities, and surgical management.

Methods: We report a 20-year retrospective review of all foregut duplication cysts managed at our institution.

Results: Twelve patients with 13 foregut duplication cysts were identified. The ages of the children at the time of surgery ranged from infancy to adolescence, with a mean age of 7.2 years. Half of the patients presented with abdominal pain and vomiting, and the remaining either had respiratory distress or were asymptomatic. All resections were performed electively. Two of the 11 patients had other congenital anomalies, including a congenital pulmonary airway malformation and coarctation of the aorta. One patient had prenatal diagnosis by ultrasonography. Nine patients underwent complete successful excision with no complications. Three patients whose symptoms resolved during hospitalization remained under observation because of parental preference.

Conclusions: Foregut malformation in children may present with a variety of symptoms or can be found incidentally. The decision and timing of surgery is based on the clinical presentation. Surgical intervention in asymptomatic patients should be based on a thorough discussion with the parents.

Key Words: Bronchogenic, Esophageal, Foregut duplication cyst, Gastric.

INTRODUCTION

Alimentary tract duplication is a relatively rare congenital anomaly. It can be found anywhere from the mouth to the anus and can be symptomatic or are discovered incidentally. W. E. Ladd first introduced the term duplication in 1934.¹ Most duplications are benign, but the presence of ectopic gastric mucosa and the potential for malignant degeneration remain a concern.²

Congenital duplication can occur anywhere in the gastrointestinal (GI) tract, although it most commonly occurs in the ileum, esophagus, and colon. One third of all duplications arise from the foregut (esophagus, stomach, and first and second part of the duodenum).³ Enteric duplication varies widely in size and is usually single, more often spherical than tubular, and lined with alimentary tract mucosa. Foregut duplication is more common in girls, particularly if there is bronchopulmonary involvement.⁴ Some duplications are identified incidentally but many cause problems in early childhood. Respiratory symptoms are common in foregut duplication, especially when there is involvement of the bronchial tree. In some cases, the patient may present with respiratory distress and hemoptysis.⁵

Because of the infrequency of foregut duplication, there has been limited analysis of clinical characteristics, presentation, and preferred mode of management. Our review was undertaken to analyze some of these features.

METHODS

Twelve patients (7 boys and 5 girls) with foregut duplication treated at the Maria Fareri Children's Hospital from 1997 through 2016 were identified from medical records (**Table 1**). Four of the foregut duplications involved the esophagus, and 5 involved the stomach. Three patients had thoracic cysts located in the posterior mediastinum. All 12 duplications were cystic. The patients' ages at surgery were between 2 and 12 years, with a mean age of 7.2 years. All patients were compliant with follow-up. Two with duplication cysts found incidentally were asymptomatic at the time of diagnosis. Three patients did not undergo surgery. These patients were symptomatic at diagnosis and showed resolution of symptoms during

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Table 1.
Characteristics of the Patients

Age at Diagnosis	Sex	Location	Presentation	Associated Findings	Diagnosis
5 years	F	Gastric: greater curvature	Vomiting and abdominal pain	None	CT Abdomen
2 years	M	Gastric: extending through GE junction sharing a common wall with esophagus	Asymptomatic ^b	Bilateral fetal caliectasis	Antenatal USG
10 years	F	Gastric-GE junction	Abdominal pain	Bilateral Inguinal Hernia, Hiatal Hernia	MRI abdomen
9 years	M	Gastric: lesser gastric curvature (2 cysts)	Vomiting and abdominal pain	None	MRI abdomen
12 years	M	Gastric: level of fundus	Abdominal pain	None	CT abdomen
3 years	M	Esophagus: distal	Respiratory distress	None	Chest x-ray
17 years ^a	M	Esophagus: mid	Upper GI bleed	None	MRI abdomen
11 years	F	Esophagus: distal	Abdominal Pain	None	MRI chest
14 years ^a	F	Esophagus: distal	Abdominal Pain	None	CT abdomen
12 years	F	Right lung	Asymptomatic ^c	Coarctation of Aorta	MRI abdomen
8 months	M	Right Lung	Respiratory distress	Congenital pulmonary Airway Malformation	MRI chest
7 years ^a	M	Left hemithorax	Chest pain, fever	None	Chest x-ray

^aPatients who refused surgery. ^bIncidental finding during antenatal ultrasound. ^cIncidental finding during postoperative investigation after coarctation of aorta repair.

their hospital admission. After a thorough explanation of risks and benefits of surgery, the parents decided on nonoperative management. They are currently being followed up.

RESULTS

Patient characteristics and symptomatology varied according to the anatomic location of the duplication. All of the surgically managed cases recovered uneventfully. There were no significant postoperative complications.

The duplication types, pathology and surgical management are shown in **Table 2**.

Esophageal Duplication Cysts

The four patients diagnosed with the esophageal duplication cyst presented with symptoms including upper GI bleed, abdominal pain and respiratory distress. The 11-year-old girl had a nonenhancing cyst at the level of the distal esophagus on her chest magnetic resonance image (MRI) (**Figure 1A**). Both the 11- and 3-year-old patients

underwent elective video-assisted thoracoscopic surgery (VATS) with uneventful recovery (**Figure 1B, C**).

The 3-year-old boy underwent removal of the cyst through a left thoracoscopy with three 5 mm ports. The cyst was slightly attached to the diaphragm and was easily removed with the Harmonic Scalpel (Ethicon Inc., Somerville, New Jersey, USA).

The 11-year-old girl had a large bronchogenic cyst firmly attached to the left lower esophagus. The cyst was removed through 3 ports. Two were 5 mm and a third was 12 mm, for placement of the endostapler. The cyst was completely removed with the endostapler, with a flexible fiberoptic endoscope placed in the esophagus to serve as a stent, to confirm water tightness, and to inspect for possible injury to the esophagus. Histology of the resected esophageal cysts was consistent with a bronchogenic cyst.

The 17-year-old patient presented with upper GI bleeding and underwent an upper endoscopy, with normal findings. His GI bleed resolved with medical management. His work-up included an MRI of the chest that showed a

Table 2.
Location, Pathology, and Operative Modality

Age at Diagnosis	Location	Pathology	Surgical Management
5 years	Gastric	Gastric mucosa	Laparoscopic removal of gastric cyst duplication with partial gastrectomy
2 years	Gastric	Gastric mucosa	Laparoscopic removal of gastric cyst duplication
10 years	Gastric	Gastric mucosa	Laparoscopic removal of gastric cyst duplication
9 years	Gastric	Gastric mucosa	Laparoscopic removal of 2 gastric cyst duplications
12 years	Gastric	Bronchogenic	Laparoscopic removal of retroperitoneal cyst
3 years	Esophagus	Bronchogenic	Left video-assisted thoracoscopic removal
11 years	Esophagus	Bronchogenic	Left video-assisted thoracoscopic removal
8 months	Right lung	Bronchogenic	Thoracoscopic right lower lobectomy with thoracoscopic excision of right-side bronchogenic cyst
12 years	Right lung	Bronchogenic	Thoracoscopic enucleation of right-side bronchogenic cyst

cystic mass adjacent to the distal esophagus. A Meckel’s scan did not demonstrate gastric mucosa within the cyst. This patient opted for observation in place of surgery. The 14-year-old girl who presented with acute worsening of her chronic epigastric abdominal pain was found to have a duplication cyst anterior to the gastroesophageal (GE) junction on her abdominal computed tomographic study (CT). She underwent an upper endoscopy that was normal. Her symptoms improved during admission, and her parents opted for her to be observed in lieu of surgery. Both patients have remained well on outpatient follow-up.

Gastric Duplication Cysts

Five patients had gastric duplication cysts. The youngest was 2 years of age, and the remainder were between 5 to 12 years of age. Two of the gastric duplications were situated close to the body and greater curvature of the stomach, whereas 1 gastric duplication was located against the lesser curvature and the other between the lesser curvature and liver.

A 12-year-old boy with a gastric duplication cyst presented with a 3-week history of epigastric abdominal pain and constipation. Abdominal CT showed a retroperitoneal cyst found abutting the greater curvature of the stomach and pancreatic body and indenting the left adrenal gland, with no involvement of the pancreatic duct. Endoscopic ultrasonography demonstrated a thin-walled cyst between

the pancreas, stomach, and adrenals. The patient underwent successful laparoscopic removal of the cyst. Pathology was consistent with a bronchogenic cyst.

The 9-year-old patient was found to have 2 communicating cysts between the gastric lesser curvature and the stomach (**Figure 2A**) as identified on the sonogram (**Figure 2B**).

The 5-year-old girl had a 5-cm gastric duplication cyst in the greater gastric curvature, sharing a common wall with the stomach. The duplication was removed with an endostapler. A flexible fiberoptic scope was placed through the stomach into the duodenum to serve as a stent and to confirm complete removal of the common gastric wall. The cyst being 5 cm in size, was accompanied by a partial gastrectomy. The staple line was not reinforced with sutures.

The 10-year-old girl with a duplication cyst at the level of the GE junction presented with upper abdominal pain. Her abdominal MRI showed a nonenhancing cystic lesion at the level of the GE junction. Based on this finding, a gastric duplication cyst was considered the most likely possibility (**Figure 3A**). She underwent successful laparoscopic excision of the cyst (**Figure 3B**).

The 12-year-old girl had a large cyst adjacent to the gastric fundus and greater curvature sharing a common wall with the stomach. The cyst was removed with the Harmonic Scalpel. Reconstruction of the esophagus and stomach was performed in 2 layers. The first layer was

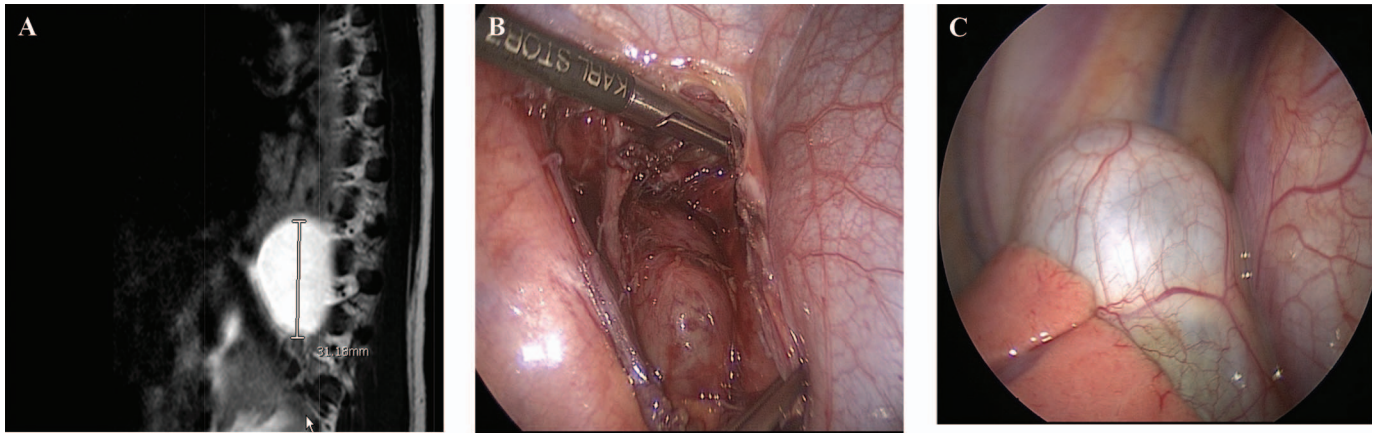


Figure 1. **A**, Chest MRI showing a cyst to the left of the esophagus overlying the T7–T8 vertebral bodies. **B, C**, Thoracoscopic view of esophageal duplication cysts.

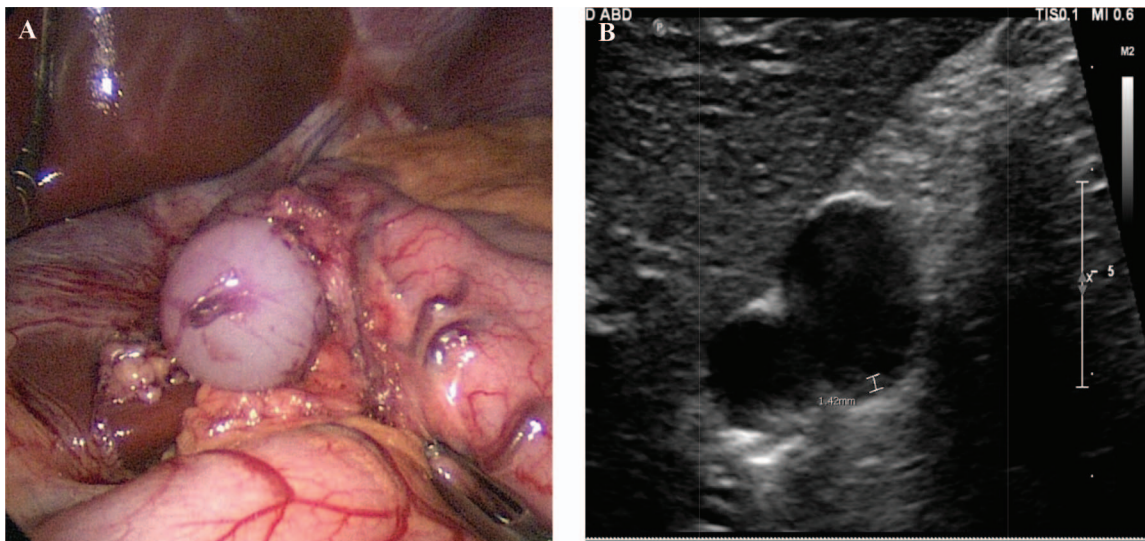


Figure 2. **A**, Laparoscopic view of two duplication cysts against the lesser curvature of the stomach. **B**, Bilobed cyst adjacent to the gastric fundus on ultrasonography.

a running suture with polyglycolic-acid sutures. A second inverting layer was applied with interrupted 2-0 silk sutures.

At the time of this writing, all 5 patients were doing well on follow up.

Thoracic Duplication Cysts

All 3 duplication cysts were located in the posterior mediastinum. The youngest patient was an 8-month-old full-term boy who was found to have a cystic chest mass on a 34-week prenatal sonogram. He was admitted to the neonatal intensive care unit because of respiratory distress

and was found to have right-side pneumothorax. An MRI of the chest revealed a ruptured 4×4 -cm large congenital cystic adenomatoid malformation in the right lower lung lobe and a 2-cm bronchogenic cyst in the right posterior lateral lung (**Figure 4**). At 8 months of age, he underwent a right lower lobe thoracoscopic lobectomy with excision of the bronchogenic cyst at the same time. Microscopically, the cyst was lined with ciliated pseudostratified columnar epithelium. His postoperative course was complicated by right-side atelectasis which resolved with pulmonary toileting and chest physiotherapy. His follow-up chest film showed good aeration of the lungs, and he was discharged on the sixth postoperative day.

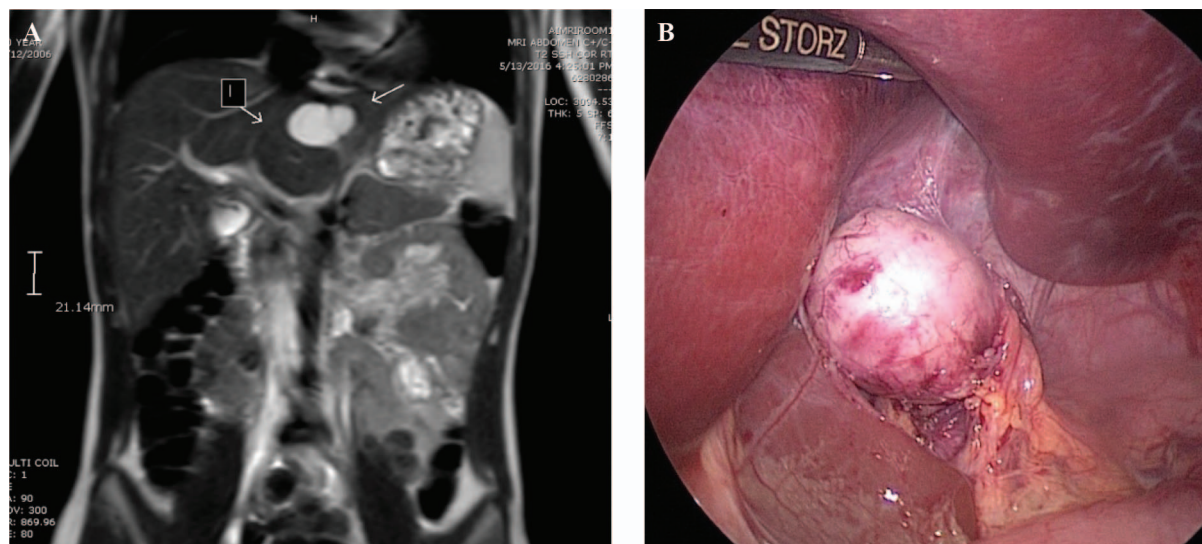


Figure 3. **A**, Abdominal MRI showing a nonenhancing cyst at the level of the GE junction. **B**, Laparoscopic view of a gastric duplication cyst.

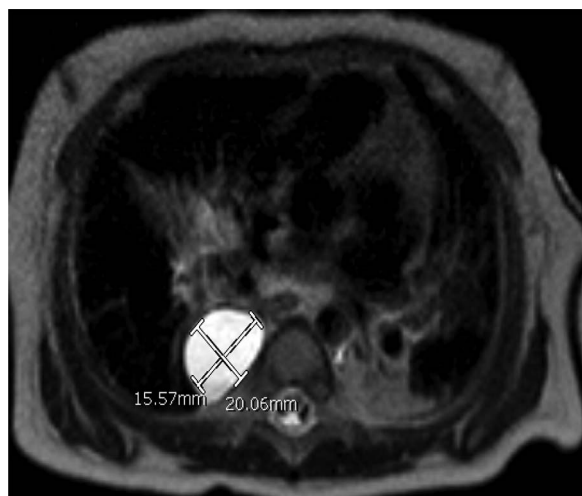


Figure 4. Chest MRI. T₂ axial image demonstrating 2-cm hyperintense cystic lesion within the right posterior lateral lung.

The second patient with a bronchogenic cyst was a 12-year-old girl who had coarctation of aorta and had undergone multiple balloon dilatations. An MRI of the chest to evaluate her aorta showed an incidental finding of a 3-cm cyst at the level of the right main stem bronchus in front of the pulmonary artery. The cyst was completely removed by VATS with three 5-mm ports and use of the Harmonic Scalpel. Histologic examination showed that both cysts had pseudostratified ciliated columnar epithelium.

DISCUSSION

Embryologically, the foregut at the cranial end of the primitive gut develops into the pharynx, respiratory tract, esophagus, stomach, and the first part and proximal half of the second part of the duodenum.⁶

Duplications are congenital malformations thought to arise from disturbances in embryologic development. Multiple theories have been postulated to account for their development. Bentley and Smith⁷ proposed that the primary defect was the development of a split notochord that allowed the connection between the yolk sac endoderm and the ectoderm and that subsequent duplication of the gut resulted from eventration of the yolk sac between the halves of the vertebra. No single theory has explained the origin, various locations involved, and the associated anomalies of the duplication cyst.

Parker et al⁸ defined duplication cyst in 1972. They described duplication of the alimentary tract to form a cystic or spherical structure attached to a part of the bowel, sharing a wall of smooth muscle and lined by a mucous membrane similar to some part of the alimentary canal. The associated findings of vertebral, spinal cord, and genitourinary malformations, as well as malrotation and intestinal atresia suggest a multifactorial process in the development of alimentary tract duplications.⁸ All of the duplication cysts in our series were cystic.

Duplication cysts usually share a common smooth muscle wall and blood supply.⁹ The symptoms are related to size, location, type of duplication, and presence of heterotopic mucosa. Alimentary tract duplications present with a wide range of symptoms including abdominal distension, abdominal pain, obstruction, bleeding, respiratory compromise, or a painless mass. Most (80%) present before 2 years of age; prenatal ultrasonography can detect duplications as early as 16 weeks of gestational age.⁸ In our series, 1 patient was diagnosed by prenatal ultrasonography.

Esophageal duplication is the second most common type after ileal duplication. Duplication of the cervical portion accounts for 23% of esophageal duplications. They usually present as enlarging neck masses or with upper airway obstructive symptoms, or they can be asymptomatic. Duplication cysts involving the mid esophagus constitute 17% of esophageal duplications. They often present with respiratory distress because of airway obstruction. Most esophageal duplications involve the distal esophagus comprising 60% of all esophageal duplications. In this position, they usually go undetected for several years, as they are often asymptomatic in this location.⁸

Esophageal duplication cyst may remain undetected on plain chest radiograph. Noncontrast and contrast-enhanced CT scan can demonstrate relation of the duplication cyst with the esophagus and tracheobronchial tree. The mainstay of treatment is surgical excision, which can be accomplished by means of a thoracotomy or thoracoscopy.

Gastric duplication presents commonly with recurrent abdominal pain, vomiting, feeding difficulty, and a palpable mass. Peptic ulceration with hemorrhage or perforation may occur if an intraluminal connection is present or if it contains ectopic gastric mucosa. Recommended management is complete excision of the duplication, irrespective of symptoms. It is preferable to remove the duplication without violating the bowel lumen. However, a large duplication may require partial gastrectomy or mucosal stripping.

Bronchogenic cyst originates from abnormal branching of the tracheobronchial anlage, which may create closed spaces lined by either esophageal or bronchial mucosa. The cyst, located close to the trachea and main-stem bronchi, is lined with ciliated epithelium.¹⁰ It can become symptomatic as a result of secreting mucosa, which progressively enlarge it. In rare instances, the foregut malformation is more extensive, involves both the respiratory and digestive tracts, and combines esophageal duplication

with airway malformation.¹¹ Bronchogenic cyst often occurs around the carina. It can be attached to, but not communicate with, the tracheobronchial tree. Although bronchogenic cyst frequently occurs in the mediastinum, it may also occur in the retroperitoneum. Most of the reported cases have been diagnosed incidentally.¹² One patient in our series was found to have a bronchogenic cyst in the retroperitoneum.

Plain chest radiographs may show paramediastinal round opacities. CT and MRI will demonstrate the size and location of the cyst and the nature of the contents. Endoscopy may be useful when the cyst is within the wall of the esophagus, and it can demonstrate external compression in some cases. Bronchogenic cysts can be removed via thoracotomy or thoracoscopy which has become the preferred approach.¹³ We recommend complete excision to confirm the diagnosis, relieve symptoms, and prevent complications. As in other duplications, it is essential to completely remove the secreting mucosa to prevent recurrence or cancer in adulthood.

In summary, foregut duplication can have a variety of clinical presentations or can be asymptomatic and found incidentally. We recommend complete surgical excision when possible. If complete removal is not feasible, then all ectopic or heterotopic tissue should be removed, including the lining of the duplication.

All of our patients were successfully treated with laparoscopy and thoracoscopy. We recommend the use of these minimally invasive techniques whenever possible. Long-term follow-up is essential when surgical intervention is refused.

References:

1. Ladd WE. Duplications of the alimentary tract. *South Med J.* 1934;30:363–371.
2. Johnson JA 3rd, Poole GV. Ileal duplications in adults: presentation and treatment. *Arch Surg.* 1994;129:659–661.
3. Hocking M, Young DG. Duplications of the alimentary tract. *Br J Surg.* 1981;68:92–96.
4. Ildstad ST, Tollerud DJ, Weiss RG, Ryan DP, McGowan MA, Martin MW. Duplications of the alimentary tract: clinical characteristics, preferred treatment and associated malformations. *Ann Surg.* 1988;208:184–189.
5. Burgner DP, Carachi R, Beattie TJ. A thoracic foregut duplication cyst presenting with neonatal respiratory distress and haemoptysis. *Thorax.* 1994;49:287–288.

6. Bremer JL. Diverticula and duplications of the intestinal tract. *Arch Pathol*. 1944;38:132–140.
7. Bentley JFR, Smith JR. Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome. *Arch Dis Child*. 1960;35:76–86.
8. Parker BC, Guthrie J, France NE, Atwell JD. Gastric duplications in infancy. *J Pediatr Surg*. 1972;7:294–298.
9. Stringer MD, Spitz L, Abel R, et al. Management of alimentary tract duplication in children. *Br J Surg* 1995;82:74–78.
10. Ribet ME, Copin MC, Gosselin B. Bronchogenic cysts of the mediastinum. *J Thorac Cardiovasc Surg*. 1995;109:1003–1010.
11. Kitano Y, Iwanaka T, Tsuchida Y, et al. Esophageal duplication cyst associated with pulmonary cystic malformations. *J Pediatr Surg*. 1995;30:1724–1727.
12. McCrystal DJ, Borzi PA. Retroperitoneoscopic resection of retroperitoneal bronchogenic cysts. *Pediatr Surg Int*. 2002;8:375–377.
13. Kawahara H, Kamata S, Nose K, et al. Congenital mediastinal cystic abnormalities detected in utero: report of two cases. *J Pediatr Gastroenterol Nutr* 2001;33:202–205.