Comment

This case is unique in the use of minimally invasive techniques to manage a spontaneous esophageal perforation. Rather than perform thoracotomy, we elected to seal the esophageal leak by placing a self-expanding covered Polyflex stent with simultaneous video-thoracoscopic drainage and debridement.¹ The morbidity of the procedures was minimal and the patient's overall condition stabilized rapidly, allowing return to oral nutrition.

Traditionally, identification of the perforation site at thoracotomy with debridement of nonviable tissue is necessary before a buttressed repair with wide drainage.² The success in the management of esophageal perforations depends on the time interval to intervention, the cause and the site of the perforation, and control of mediastinal contamination and restoration of esophageal continuity.³

This case illustrates unique management of a spontaneous esophageal perforation by combining minimally invasive video-assisted thoracoscopic surgery with use of a flexible endoscope. Thoracoscopic drainage and debridement with upper endoscopy placement of a Polyflex stent sealed the esophageal perforation.

The patient did well and eventually returned for outpatient stent removal.

Spontaneous esophageal perforation may not require thoracotomy in all cases. In cases in which no underlying esophageal disease exists, a combination of upper endoscopy and thoracoscopy may allow adequate management of this patient population.⁴

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Dextrocardia is a component of left-sided Poland syndrome

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The combination of Poland syndrome and dextrocardia is very uncommon, only 17 cases having been published until now. In all of them, the manifestations of Poland syndrome were on the left, although they usually occur on the right. We report the case of left-sided Poland syndrome combined with isolated dextrocardia, bilateral palmar hyperhydrosis, and hypoplasia of the rectus abdominis muscle. Clinical correlations between Poland syndrome and dextrocardia are discussed.

Clinical Summary

A 20-year-old man who was admitted to our hospital had previously been given a diagnosis of Poland syndrome. There were no relevant anomalies in the family. On examination, there was a defect in the left pectoralis major and minor muscle

attachments to the sternum through which the skin retracted with respiration. The left areola was small and located higher than the right one, with an inverted nipple (Figure 1). Hypoplasia of subcutaneous tissue, alopecia of the mammary and axillary regions, and bilateral palmar hyperhydrosis were detected. A posteroanterior view of the patient showed isolated dextrocardia, absence of the left costosternal portion of the pectoralis major and minor muscles, and deformity of the costal cartilages of the second, third, and fourth ribs on the left (Figure 2). Echocardiography demonstrated isolated dextrocardia and no evidence of intracardiac or great vessel anomaly. Hypoplasia of the rectus abdominis muscle was noted on abdominal ultrasound. There were no signs of digital anomalies or Möbius syndrome. The karyotype appeared normal.

Discussion

Poland's syndrome is a rare congenital anomaly characterized by hypoplasia or absence of the breast or nipple, hypoplasia of subcutaneous tissue, absence of the costosternal portion of the pectoralis major muscle, absence of the pectoralis minor muscle, aplasia or deformity of costal cartilages of ribs 2, 3, and 4 or 3, 4, and 5, alopecia of the mammary and axillary region, and ipsilateral hand deformities. Clinical manifestations of Poland syndrome are extremely variable, and rarely are all the features recognized in one individual. Depending on the physician's specialty and the referral pattern, a variable incidence of the anomalous defects is recognized. In some cases other associated ipsilateral anomalies (dextrocardia, vertebral defects, renal

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Received for publication May 18, 2005; accepted for publication June 30, 2005.

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J Thorac Cardiovasc Surg 2005;130:1471-2

0022-5223/\$30.00

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doi:10.1016/j.jtcvs.2005.06.048

Figure 1. Anterior aspect of chest wall.

Figure 2. Chest radiograph showing isolated dextrocardia and deformity of the left second, third, and fourth ribs.

aplasia or hypoplasia, undescended testes, Möbius syndrome, lung herniation, tumors) have been described. The incidence of Poland syndrome, reported by different authors, ranges from 1:10,000 to 1:100,000. The syndrome is observed more frequently in male than in female subjects, with the right side of the body affected more often than the left.⁵ The etiology of this syndrome is still under discussion. However, although most described cases were sporadic, rare familial instances of Poland syndrome have also been presented. Different etiologic factors of Poland syndrome are taken into account: genetic, vascular compromise during early stages of embryogenesis, but also teratogenic effects of environmental xenobiotics.

Situs inversus totalis is defined as complete mirror-image transposition of the abdominal and thoracic viscera, occurring during development.³ Isolated dextrocardia (*situs solitus*) represents an anomaly with normal situs but a right-sided heart. It also means that both the abdominal viscera and the atria are in the normal position. Radiographically, *situs solitus* is a certainty when both the aortic knob and the gastric air bubble are on the left side. The estimated incidence of isolated dextrocardia varies from 1 in 30,000 to 1 in 36,000 live births.^{4,5}

Isolated dextrocardia has been reported in 17 patients with Poland syndrome. In all these patients, Poland syndrome was left-sided and associated with rib defects, although its manifestations usually occur on the right.^{4,5} Dextrocardia was reported in 5.6% of a series of 144 patients with Poland syndrome, and in 9.6% of those, the defect was left-sided.^{1,4,5} In patients with isolated dextrocardia, the incidence of congenital heart disease

has been estimated at 98%. In dextrocardia with *situs inversus* this rate is only 5%.² Congenital cardiovascular anomalies have not been reported in Poland syndrome with dextrocardia. Dextrocardia in Poland syndrome is associated with rib defects in all of cases, whereas rib defects are reported in only about 15% of patients with right-sided Poland syndrome.⁴

Our case also supports the view that the combination of Poland sequence and dextrocardia is not coincidental and dextrocardia may be part of the Poland syndrome, especially left-sided. Further studies would be required to confirm this unusual combination.

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