Case Report

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Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.151000
Quick Response Code:

Unilateral pulmonary agenesis associated with oesophageal atresia and tracheoesophageal fistula: A case report with prenatal diagnosis

Go Miyano, Keiichi Morita, Masakatsu Kaneshiro, Hiromu Miyake, Mariko Koyama, Hiroshi Nouso, Masaya Yamoto, Reiji Nakano[1], Yasuhiko Tanaka[1], Tomizo Nishiguchi[2], Takakazu Kawamura[2], Koji Fukumoto, Naoto Urushihara

ABSTRACT

We describe herein a case of unilateral pulmonary agenesis (PA) with oesophageal atresia (EA)/tracheoesophageal fistula (TEF) that was diagnosed prenatally and repaired by esophagoesophagostomy with stable postoperative course. The patient was born at 34 weeks gestation, after ultrasonography at 22 weeks gestation showed possible right-sided diaphragmatic eventration or PA and EA was subsequently suspected due to hydramnios. The initial X-ray showed mediastinal shift to the right, and coil up sign of the nasogastric tube, without intracardiac anomaly. Immediately after the diagnosis of EA/TEF and unilateral PA on day 0, the patient was intubated in the operating room, and a gastrostomy tube was placed. After pulmonary status stabilized, at 4 days old, EA/TEF was repaired through a thoracotomy in the right 4th intercostal space. The right main bronchus was noted to continue into the distal oesophagus; this fistula was ligated and divided, and a single-layer esophagoesophagostomy was performed under mild tension with one vertebral gap. The neonate was maintained on mechanical ventilation and gradually weaned to extubation at 7 days old. The postoperative course was uneventful, with the exception of prolonged jaundice that emerged at 3 months old. Laparoscopic cholangiography at that time excluded biliary atresia, and jaundice resolved spontaneously. The patient has not shown any respiratory symptoms or feeding difficulties as of the 12-month follow-up.

Key words: Oesophageal atresia, tracheoesophageal fistula, unilateral pulmonary agenesis

Departments of Pediatric Surgery, [1]Neonatology and [2]Obsterics, Shizuoka Children's Hospital, Shizuoka, Japan

Address for correspondence:
Dr. Go Miyano,
Department of Pediatric Surgery, Shizuoka Children's Hospital, 860 Urushiyama, Aoi-ku, Shizuoka 420-8660, Japan.
E-mail: go1993@hotmail.co.jp

INTRODUCTION

The association of unilateral pulmonary agenesis (PA) with oesophageal atresia (EA) and tracheoesophageal fistula (TEF) is exceedingly rare, and this combination of anomalies has been uniformly lethal. We describe a case of unilateral PA with EA/TEF that was diagnosed prenatally and repaired by esophagoesophagostomy with stable postoperative course.

CASE REPORT

The patient was born at 34 weeks gestation to a 31-year-old mother after ultrasonography at 22 weeks gestation showed possible right-sided diaphragmatic eventration or PA. EA was subsequently suspected due to hydramnios. No prenatal chromosomal analysis was documented. At an estimated gestational age of 31 weeks, when poor heart rate variability and late decelerations developed in the foetus, emergency caesarean section was performed. A male baby was born with an anomalous birth weight of 1370 g. The initial X-ray showed a mediastinal shift to the right and coil up sign of the nasogastric tube [Figure 1]. Both findings were indicative of possible EA/TEF, and of a collapsed or absent right lung. Echocardiography showed that the heart was located in the right thorax, with no intracardiac anomaly. Immediately after the diagnosis of EA/TEF and unilateral PA on day 0, the patient was intubated in the operating room, and a gastrostomy tube was placed without any complications related to TEF, like aspiration pneumonitis. After pulmonary status had stabilized, at 4 days old, the EA/TEF was repaired through a thoracotomy in the right 4th intercostal space. On right posterolateral thoracotomy, the pericardium was seen to be immediately deep to the ribs, and no right lung was evident [Figure 2]. The distal oesophagus was

traced upward to its communication with the trachea. The tracheal bifurcation was identified, and the right main bronchus was noted to continue into the distal oesophagus; this fistula was ligated and divided. A single-layer esophagoesophagostomy was performed under mild tension with 1 vertebral gap. The neonate was maintained on mechanical ventilation and gradually weaned to extubation at 7 days old. A water-soluble contrast study on postoperative day 5 revealed no leakage [Figure 3], and the patient advanced to full oral feeding by 7 days old. The postoperative course was uneventful, with the exception of prolonged jaundice that emerged at around 1-month-old with grey stool and T- and D-Bil levels of 3.6 and 2.8 mg/dl, respectively. Laparoscopic cholangiography at 2-month-old excluded the possibility of biliary atresia with clear images of the intrahepatic bile duct, and symptoms resolved spontaneously with conservative management. The patient was discharged to home at 3 months old. He has been tolerating soft foods orally and gaining weight without supplemental gastrostomy feedings, so the gastrostomy was closed. The patient has not shown any respiratory symptoms or feeding difficulties for 12 months. Developmental testing shows normal intelligence without any delay in motor ability.

DISCUSSION

More than 50% of patients with PA have associated anomalies, and 50% of children born with PA are stillborn or die within the 1st months of life from these concomitant anomalies.[1] Unilateral PA associated with EA-TEF is a rare condition, and according to a review of the literature, although survival has been reported in a very small number of cases, most cases of mortality died of associated anomalies, such as congenital cardiac disease or refractory pulmonary problems.[1] However, there seems to be a clear tendency that patients with EA and TEF and associated anomalies related to congestion, such as duodenal atresia and hypertrophied pyloric stenosis, show relatively good prognosis. The prognosis tends to depend on the presence of associated cardiac or pulmonary anomalies. Although our patient experienced prolonged jaundice warranting laparoscopic cholangiography, he has followed a stable postoperative course for more than a year after surgery. The use of foetal ultrasonography has significantly improved the ability to diagnose foetal thoracic disease.[2] Abnormal findings such as mediastinal shift and the presence of overwhelmingly unilateral pulmonary parenchyma in the foetal hemithorax may include additional pathologies of PA other than congenital diaphragmatic hernia, eventration, or dextrocardia, necessitating proper follow-up during

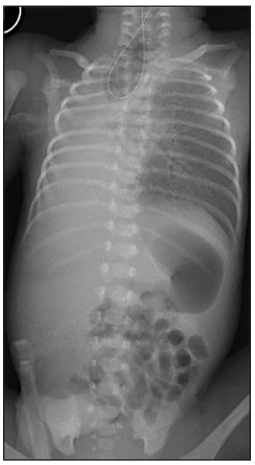

Figure 1: Chest X-ray on day 0. Mediastinal shift to the right and coil up sign of the nasogastric tube are shown

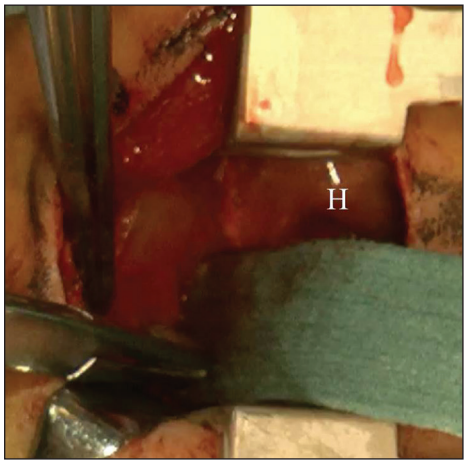

Figure 2: Intraoperative view through the thoracotomy. The pericardium is immediately deep to the ribs, and no right lung is apparent. H: Heart

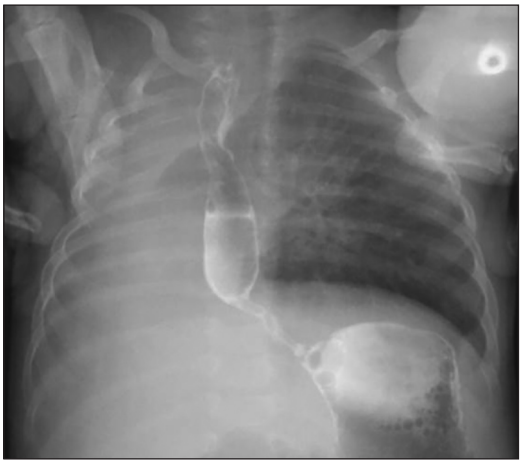

Figure 3: Postoperative esophagography on postoperative day 5. No leakage is revealed, and acceptable passage of esophagoesophagostomy is confirmed

pregnancy. However, the lack of pulmonary reserve in these patients should not be underestimated, and all efforts must be made to avoid even minor episodes of aspiration.[3] Due to the possibility of prenatally diagnosed PA and EA, we were able to make an accurate diagnosis and start the initial treatment immediately after diagnosis, and a gastrostomy was placed immediately after the respiratory status was stabilized. Thus, as the ability to achieve prenatal diagnosis becomes more common and better developed, initial postnatal intensive treatment will become more substantial and will likely to contribute to improvements in prognosis for such patients.

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Cite this article as: Miyano G, Morita K, Kaneshiro M, Miyake H, Koyama M, Nouso H, *et al.* Unilateral pulmonary agenesis associated with oesophageal atresia and tracheoesophageal fistula: A case report with prenatal diagnosis. Afr J Paediatr Surg 2015;12:86-8.

Source of Support: Nil. **Conflict of Interest:** None Conflict of Interest.