check up and BA was diagnosed. The hepatic portojejunostomy (Kasai's procedure) was performed at the age of 135 days. Subsequently, ascending cholangitis occurred frequently and administration of antibiotics was required.

In April 2000, TB rose above 1 mg/dL. Ascites appeared in July 2002 and liver function deteriorated (albumin: 2.9 g/dL, AST: 120 IU/L, ALT: 91 IU/L, TB: 10.2 mg/dL), despite water restriction and diuretics. She was referred to our hospital for LDLT in May 2003.

The problem was that before transplantation, her white blood cell count ranged from 3000/mm^3 to 39 000/mm^3 with 60–95% eosinophils. Mild eosinophilia was noted around October 2001, ranging from 10 to 15%, but the patient did not have any symptoms other than mild liver dysfunction supposed to be due to BA, so we did not give specific treatments. Then eonosinophil count gradually rose up to 95% (Fig. 1). Immunoglobulin E was 3260 mg/dL and vitamin B12 was 4160 pg/mL at that time.

To rule out drug-induced eosinophilia, drug lymphocyte-stimulating test was performed for H-2 blockers, penicillin, and cephalosporine. Results were all negative against these drugs. She had a history of asthma with only minor respiratory symptoms, but had no other allergies including foods or symptoms of vasculitis associated with Churg-Strauss vasculitis. Anti-nuclear antibody, anti-mitochondrial antibody, and perinuclear antineutrophil cytoplasmatic antibody were negative. Antibody tests and stools were negative for ova and parasites. Bone marrow biopsy showed elevated eosinophils with no evidence of malignancy (NCC: 385 000/mm^3, eosino: 58.0%, normal karyotype), ECG and chest X-ray findings were normal. An abdominal ultrasound examination showed hepatosplenomegaly, dilatation and thickening of the intesitinal wall and ascites. Upper and lower gastrointestinal tract endoscopy revealed edema of the gastric and the rectal mucosa, the biopsy specimens showed mature eosinophilic infiltration. Esophageal varices (Li, F1, Cb, RC-) were also observed.

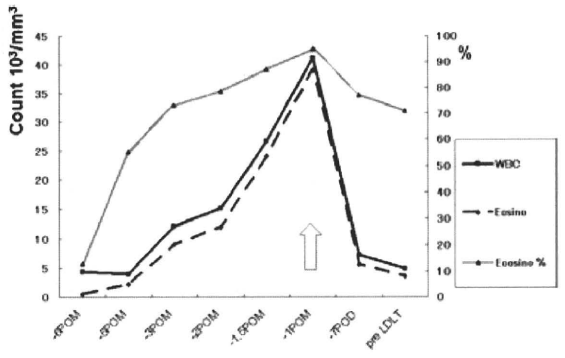

Fig. 1. Preoperative peripheral eosinophil count (arrow indicates the timing of administration of the antibiotic because of acute cholangitis). POM, preoperative months; POD, preoperative days.

We were concerned that hypereosinophilia would have a negative effect on the management of LDLT perioperatively. However, we decided to perform LDLT for this patient, because there was no serious organ dysfunction and we believed that strong perioperative immunosuppressive therapy would definitely be effective for hypereosinophilia. As this case was ABO blood type incompatible, we introduced our original protocol consisting of plasma exchange, splenectomy, triple immunosuppressive regimen, and intraportal infusion therapy (1). Furthermore, antibiotics administered for cholangitis

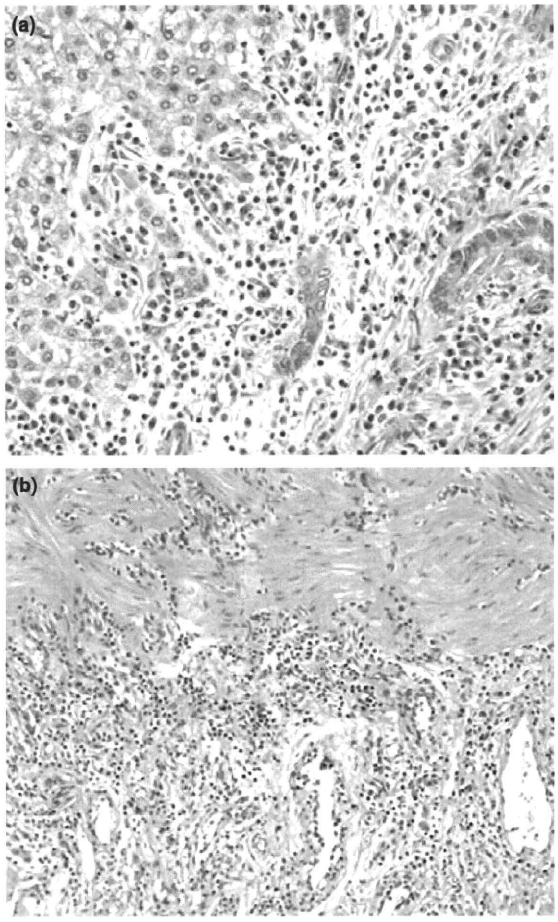

Fig. 2. (a) The resected liver: biliary cirrhosis with dense eosinophilic infiltration of portal tracts and the lobules of the liver. (b) The resected small intestine: eosinophilic infiltrations were found in the serous membrane and muscular layer of the small intestine.

preoperatively lowered the eosinophil count, from 39 000 (95%) to 4800/mm^3 (70%) (Fig. 1). In addition to this episode, the liver biopsy several months before transplantation, eosinophil count was less than 500/mm^3 at that time, showed liver cirrhosis mainly due to cholestatic liver disease. Therefore, steroids were not administered prior to transplantation. In 2004, LDLT was performed. Laparotomy showed a firm liver compatible with cirrhosis. The gastric and intestinal walls were markedly thickened, suggesting a motility disorder. Histological examination of the resected liver showed biliary cirrhosis with dense mature eosinophilic infiltration of portal tracts and the lobules of the liver, and the fibrous thickening of the inner membrane of the portal veins containing organized thrombi with abundant eosinophils (Fig. 2a). In addition, mature eosinophilic infiltration was found in the spleen as well as the serous membrane and muscular layer of the small intestine (Fig. 2b).

On post-operative day 10, she presented with fever and purulent discharge was obtained from the drainage tube (Fig. 3a). Abdominal CT was performed. The findings revealed portal vein thrombosis, periportal edema, and fluid collection at the subphrenic area (Fig. 3b). Thrombectomy and drainage were performed immediately. The histopathological findings of the thrombus revealed dense eosinophilic deposition on the organized thrombus, suggesting that HES might have influenced the formation of the thrombus (Fig. 3c). After thrombectomy, she was given heparin (5 U/kg/h) continuously and there was no thrombosis recurrence.

On post-operative day 23, because TB rose above 15 mg/dL, liver biopsy was performed. Blood chemistry findings were as follows (AST: 114 IU/L, ALT: 187 IU/L). The biopsy specimen taken of the graft demonstrated the presence of activated eosinophilils in the portal area and venulitis with eosinophilic deposition under the inner membrane, a large number of acidophilic bodies and mild inflammation in lobular area (Fig. 4). Based on those findings, although the differential count of eosinophil was less than 5% of leukocytes, HES might have caused these changes in the liver graft. Methylprednisolone (2 mg/kg/day) improved the liver dysfunction.

Currently, four yr after LDLT, despite several episodes of eosinophilic infiltration in the liver requiring extra oral steroids (2 mg/kg/day) followed by gradual dose reduction, the patient remains clinically well, on prednisolone at 0.3 mg/kg/day, with an eosinophil count ranging from 10 to 15%.

Discussion

No case of BA coexisting with HES has been reported previously. The criteria for idiopathic HES are as follows (2, 3):

- Blood eosinophilia of >1500/mm^3 is present on at least two occasions;

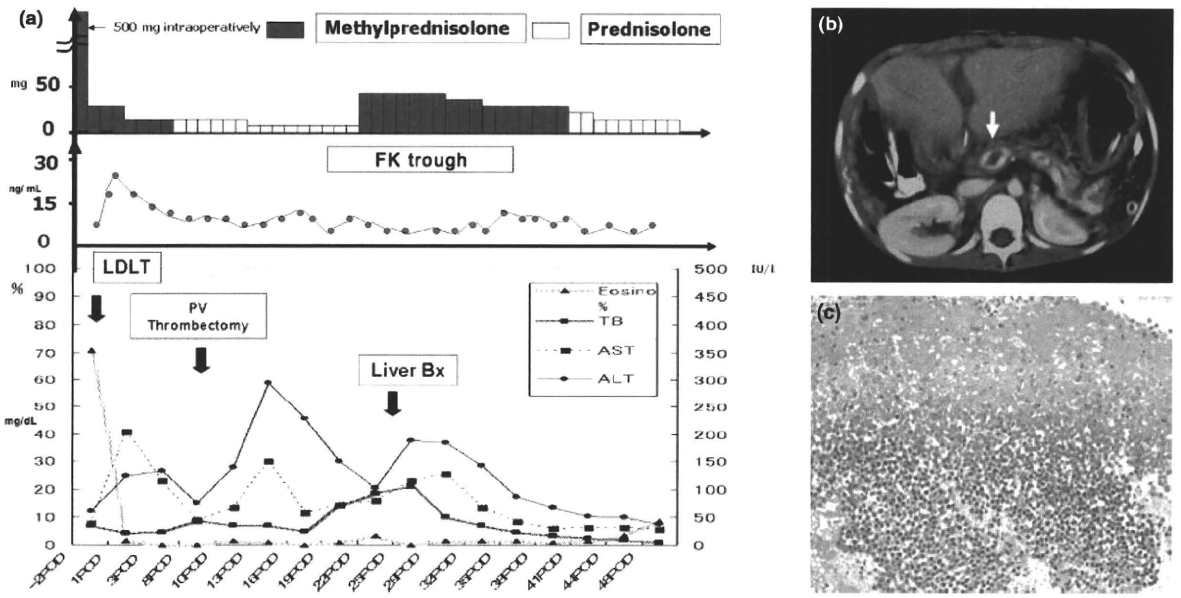

Fig. 3. (a) Clinical course. (b) CT on post-operative day 10. CT revealed portal vein thrombosis, emergent thrombectomy was performed. (c) Histology of thrombus revealed dense eosinophilic deposition on the organized thrombus.

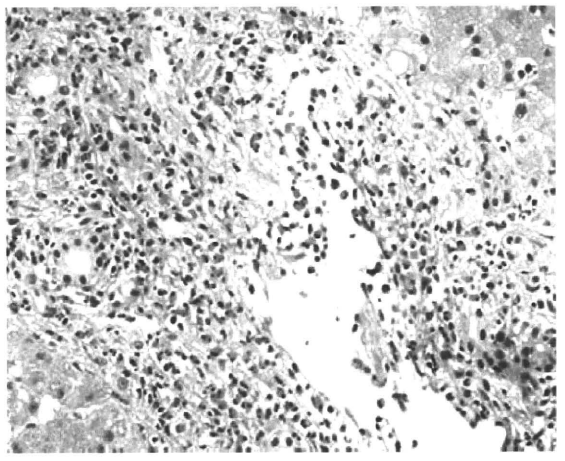

Fig. 4. The liver graft biopsy on post-operative day 23. The liver graft biopsy demonstrated the presence of activated eosinophilils in the portal area with biliary damage and venulitis with eosinophilic deposition under the inner membrane, suggesting interface hepatitis with infiltration of eosinophils. Many acidophilic bodies and mild inflammation were observed in lobular area.

- No other apparent etiologies for this degree of eosinophilia.

In this patient, HES was ultimately diagnosed post-operatively utilizing the pathological findings of the resected liver and intestine. Before the LDLT, we thought that the main reason of the liver cirrhosis was cholestasis as a result of BA. Therefore, we performed LDLT without administering steroids preoperatively. If a liver biopsy before the LDLT had shown pathological findings mainly caused by HES, we would have used corticosteroids and could have postponed the LDLT.

HES occurs more commonly in men than women, with a ratio of 9:1. Most patients are diagnosed between 20 and 50 yr of age, although HES can develop in children (4). The onset of HES is often insidious and eosinophilia may be detected incidentally as seen in this case. The underlying pathogenic mechanism for HES remains controversial. Overproduction of eosinophilopoetic signals, such as IL3, IL5, and GM-CSF, possibly secondary to abnormalities in the T-cell clones which normally produce these molecules, abnormalities of the eosinophilopoietic cytokines *per se*, defects in the receptors for eosinophilopoietic signals or in signal transduction from receptors that mediate eosinophilopoiesis, have all been proposed (2, 5).

It is interesting to know the mechanism by which HES could influence the course of the patient with BA following LDLT. Liver involvement is reported to be 20–30% in patients with HES showing hepatomegaly and minor abnormalities on liver function tests (6). The spectrum of pathologic findings including congested sinusoids, chronic active hepatitis without cirrhosis, and periportal inflammation, focal hepatic lesions, and eosinophilic cholangitis (7). Shimomura et al. (8) and Grauer et al. (9) reported eosinophilic cholangitis, an entity that mimicked PSC in the context of HES. Activated eosinophils are known to be associated with cell damage. Cytotoxic effects of the so-called major basic protein and other eosinophilic granule proteins have also been described. Foong et al. (10) demonstrated the presence of major basic protein in areas of hepatocyte injury in their patient with primary acute and chronic hepatitis. In our case, HES might have had an adverse effect on the course of BA and LDLT by a similar mechanism.

If the eosinophilic infiltration produces organ dysfunction, corticosteroid therapy should be initiated. In our case, intravenous methylprednisolone followed by oral prednisolone was administered for management of eosinophilic hepatitis (demonstrated on day 23 graft biopsy) of the post-transplantation course, which successfully improved liver function. Previous studies have shown that lymphokine-mediated T-lymphocyte control of human eosinopoiesis may play a role in the pathogenesis of HES. Steroids and calcineurin inhibitors were shown to inhibit formation of eosinophil colony stimulating factor by T-cell clones completely (11). In theory, the drug blocks the proliferation of T-cell clones that drive the excess eosinophilopoiesis in some patients (12). On the other hand, imbalance between type 1 helper T cells and type 2 helper T cells caused by the use of calcineurin inhibitors may worsen the eosinophila by over expression of IL5. Indeed, T cells from patients with HES have been shown to produce IL-5 after *in vitro* stimulation with IL-2 (13–15). However, the exact mechanism of eosinophilia after IL-2 activation remains to be clarified.

Thrombotic and thromboembolitic events are a major cause of morbidity among patients with HES (6, 7). The mechanism of thrombus formation in HES remains speculative. Eosinophils contain neurotoxic granule protein, eosinophil-derived neurotoxin, major basic protein, and ECP. ECP can incite thrombosis by binding heparin and neutralizing its anticoagulating property, as well as shortening the coagulation time of normal plasma via an interaction with factor XII (7, 16). In our case, we cannot rule out the possibility that splenectomy and the portal

vein catheter influenced hemodynamics and even caused thrombus formation. However, because the thrombus was composed of numerous eosinophils, it was suggested that HES played a major role in forming the portal thrombus. Although previous reports have demonstrated that there is no clear evidence to administer anti-platelet agents or warfarin as prophylactic therapies (7, 17, 18), we administered heparin for two wk after thrombectomy with no recurrence of portal vein thrombosis. Our protocol of anti-thrombotic prophylaxis consisted of continuous administration of low dose heparin (5 U/kg/h) followed by an oral anti-platelet agent for three months. Traditionally, the dosage for venous thrombosis has been adjusted six h later to prolong the aPTT to 1.5–2.5 times the control value (19), but no therapeutic range is appropriate if the patient's pretreatment aPTT is abnormal such as in patients immediately after liver transplantation. Therefore, in deciding about the dose and the duration of anticoagulation, consideration must be given not only to the eosinophil count but also to surgical problems after LDLT. Based on the previous reports and our experience, once the eosinophila is controlled, the need for continued anticoagulation should be determined using the same criteria used for patients without eosinophilia (20). If eosinophilia is controlled and image studies reveal the absence of thrombus or resolution of thrombus, it is reasonable to discontinue anticoagulation; however, the balance between the risks and benefits of anticoagulation must be reassessed continuously.

Moreover, there were several other episodes of liver dysfunction because of eosinophilic infiltration during the post-transplantation course, requiring extra oral prednisolone. It was difficult to clearly differentiate between eosinophilic infiltration and acute rejection in this patient; however, the presence of increasing number of acidophilic bodies in the lobular area was informative in deciding the treatment plan. It is essential to follow eosinophil counts as well as liver function carefully.

Conclusion

This case demonstrates the possibility of an association between eosinophilic infiltration and liver dysfunction during follow-up for BA and after liver transplantation. Liver transplantation with our protocol was successful despite ABO blood type incompatibility and HES, although the patient experienced portal vein thrombosis and several episodes of eosinophilic infiltration in the liver.

References

1. Tanabe M, Shimazu M, Wakabayashi G, et al. Intraportal infusion therapy as a novel approach to adult ABO-incompatible liver transplantation. Transplantation 2002: 73: 1959–1961.
2. Chusid MJ, Dale DC, West BC, Wolff SM. The hypereosinophilic syndrome: Analysis of fourteen cases with review of the literature. Medicine 1975: 54: 1–27.
3. Klion AD, Bochner BS, Gleich GJ, et al. Approaches to the treatment of hypereosinophilic syndromes: A workshop summary report. J Allergy Clin Immunol 2006: 117: 1292–1302.
4. Weller PF, Bubley GJ. The idiopathic hypereosinophilic syndrome. Blood 1994: 83: 2759–2779.
5. Spry CJ. The hypereosinophilic syndrome: Clinical features, laboratory findings and treatment. Allergy 1982: 37: 539–551.
6. Fauci AS, Harley JB, Roberts WC, Ferrans VJ, Gralnick HR, Bjornson BH. NIH conference. The idiopathic hypereosinophilic syndrome. Clinical, pathophysiologic, and therapeutic considerations. Ann Intern Med 1982: 97: 78–92.
7. Spry CJ, Davies J, Tai PC, Olsen EG, Oakley CM, Goodwin JF. Clinical features of fifteen patients with the hypereosinophilic syndrome. Q J Med 1983: 52: 1–22.
8. Shimomura I, Takase Y, Matsumoto S, et al. Primary sclerosing cholangitis associated with increased peripheral eosinophils and serum IgE. J Gastroenterol 1996: 31: 737–741.
9. Grauer L, Padilla VM III, Bouza L, Barkin JS. Eosinophilic sclerosing cholangitis associated with hypereosinophilic syndrome. Am J Gastroenterol 1993: 88: 1764–1769.
10. Foong A, Scholes JV, Gleich GJ, Kephart GM, Holt PR. Eosinophil-induced chronic active hepatitis in the idiopathic hypereosinophilic syndrome. Hepatology 1991: 13: 1090–1094.
11. Raghavachar A, Fleischer S, Frickhofen N, Heimpel H, Fleischer B. T lymphocyte control of human eosinophilic granulopoiesis. Clonal analysis in an idiopathic hypereosinophilic syndrome. J Immunol 1987: 139: 3753–3758.
12. Nadarajah S, Krafchik B, Roifman C, Horgan-Bell C. Treatment of hypereosinophilic syndrome in a child using cyclosporine: Implication for a primary T-cell abnormality. Pediatrics 1997: 99: 630–633.
13. Cogan E, Schandene L, Crusiaux A, Cochaux P, Velu T, Goldman M. Brief report: Clonal proliferation of type 2 helper T cells in a man with the hypereosinophilic syndrome. N Engl J Med 1994: 330: 535–538.
14. Rand TH, Silberstein DS, Kornfeld H, Weller PF. Human eosinophils express functional interleukin 2 receptors. J Clin Invest 1991: 88: 825–832.
15. Takamizawa M, Iwata T, Watanabe K, et al. Elevated production of interleukin-4 and interleukin-5 by T cells in a child with idiopathic hypereosinophilic syndrome. J Allergy Clin Immunol 1994: 93: 1076–1078.
16. Wassom DL, Loegering DA, Solley GO, et al. Elevated serum levels of the eosinophil granule major basic protein in patients with eosinophilia. J Clin Invest 1981: 67: 651–661.
17. Schulman H, Hertzog L, Zirkin H, Hertzanu Y. Cerebral sinovenous thrombosis in the idiopathic hypereosinophilic syndrome in childhood. Pediatr Radiol 1999: 29: 595–597.
18. Hanowell ST, Kim YD, Rattan V, MacNamara TE. Increased heparin requirement with hypereosinophilic syndrome. Anesthesiology 1981: 55: 450–452.
19. Hirsh J, Raschke R. Heparin and low-molecular-weight heparin: The Seventh ACCP Conference on Antithrombotic and Thrombolytic Therapy. Chest 2004: 126(3 Suppl.): 188S–203S.
20. Ogbogu PU, Rosing DR, Horne MK III. Cardiovascular manifestations of hypereosinophilic syndromes. Immunol Allergy Clin North Am 2007: 27: 457–475.

Pediatr Surg Int (2011) 27:103–106
DOI 10.1007/s00383-010-2716-0

CASE REPORT

A long-term survival case of tracheal agenesis: management for tracheoesophageal fistula and esophageal reconstruction

Yasushi Fuchimoto · Masaharu Mori · Fumika Takasato · Hirofumi Tomita · Yuki Yamamoto · Naoki Shimojima · Ken Hoshino · Goro Koinuma · Yasuhide Morikawa

Published online: 21 September 2010

Abstract Tracheal agenesis is a very rare disorder which leads to severe respiratory disorders immediately after birth. Reports are very limited on long-term survival cases. We report here a long-term survival case with Floyd's type I tracheal agenesis. During the neonatal stage, the patient underwent abdominal esophageal banding to substitute esophagus for trachea and transection at the cervical esophagus with esophagostomy. Subsequently, airway management was difficult due to a fragile tracheoesophageal fistula, but the fistula was conservatively treated and stabilized with the patient's growth. This patient is a very rare case in whom oral feeding was achieved after esophageal reconstruction using a gastric tube. For this case, we describe mainly (1) the management method of the tracheoesophageal fistula and (2) esophageal reconstruction without thoracotomy.

Keywords Tracheal agenesis · Tracheoesophageal fistula · Esophageal reconstruction · Floyd's type I · Long-term survival

Abbreviations

PEEP	Positive end expiratory pressure
IMV	Intermittent mandatory ventilation
CPAP	Continuous positive airway pressure

Y. Fuchimoto (✉) · M. Mori · F. Takasato · H. Tomita · Y. Yamamoto · N. Shimojima · K. Hoshino · Y. Morikawa
Department of Pediatric Surgery, School of Medicine, Keio University, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan
e-mail: yfuchimo@sc.itc.keio.ac.jp

G. Koinuma
Department of Pediatrics, School of Medicine, Keio University, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan

Introduction

Tracheal agenesis is rare among neonatal disorders. Its prognosis is very poor because its clinical symptoms are usually severe, and many cases die soon after birth [1–3]. We report herein a long-term survival case of Floyd's type I tracheal agenesis. The patient's esophagus was used as a tracheal substitute. Then he was provided strict conservative respiratory management for the fragile tracheoesophageal fistula and underwent esophageal reconstruction with a gastric tube. He remains alive at age 4 years 6 months.

Case report

An infant boy with a birth weight of 2,550 g was born by Cesarean section at 37 weeks and 4 days of gestation. Antenatal polyhydramnios was noted. The baby had no audible cry after birth. Respiratory stability was not achieved by mask ventilation, so he was resuscitated by intubation (later determined to be esophageal intubation and not endotracheal intubation). A transverse colostomy was performed at age 0 day for imperforate anus. When injection of milk was initiated through the gastric tube at age 4 days, the patient's respiratory condition deteriorated. At age 5 days, esophagography showed the trachea and bronchi (Fig. 1a). Bronchoscopy revealed the epiglottis but no glottis, and the distal segment in the direction of the trachea ended blindly (Fig. 1b). A fistula was observed extending from the esophagus to the trachea, and the patient was diagnosed with tracheal agenesis. Endoscopy and 3D-CT showed a tracheoesophageal fistula 5 cm distal to the esophageal inlet, and the tracheal bifurcation was approximately 1.5 cm from the fistula. The patient was diagnosed with Floyd's type I tracheal agenesis. The right

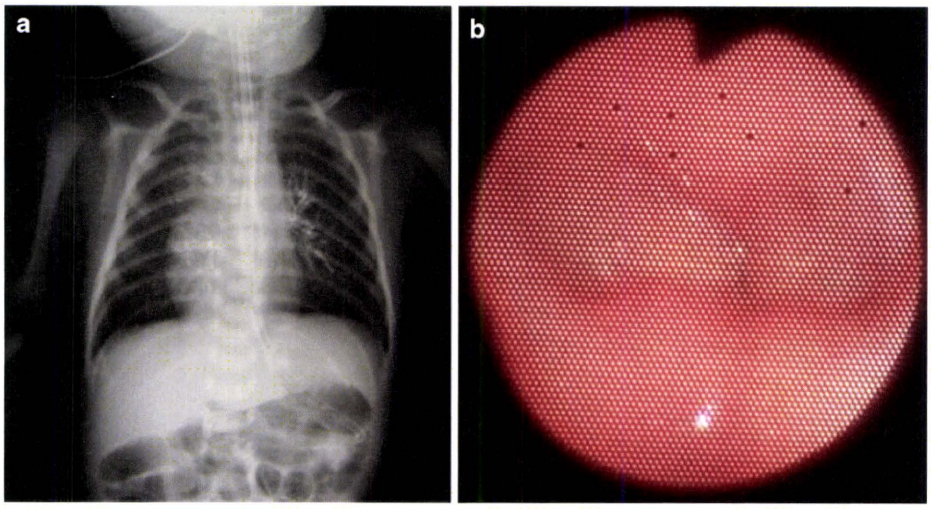

Fig. 1 **a** Esophagography at age 5 days. Contrast enhanced imaging of the esophagus showed trachea and bronchi. **b** Bronchoscopy showed the epiglottis but the distal portion ended blindly with no trachea

upper lobe bronchus arose in an area 0.8 cm from the fistula and formed a tracheal bronchus (Fig. 2). The tracheoesophageal fistula was narrow. Bronchoscopy revealed a tracheoesophageal fistula originating from the anterior esophageal wall, and the diameter of the fistula changed markedly with respiration. The fistula was a pinball size during expiration (Fig. 3a).

The patient had other malformations such as an imperforate anus, atrial septal defect, hypospadias, and polydactyly. At age 13 days, he underwent laparotomy and abdominal esophageal banding at the upper gastroesophageal junction with gastrostomy. Bronchoscopy showed that the tracheoesophageal fistula was tracheomalacia-like. In such a case, the fistula can narrow easily and tracheal secretion clearance can be impaired. Thus, expiratory impairment can frequently develop due to salivary drainage. Therefore, resection of the cervical esophagus was performed at age 53 days, and a salivary fistula was created with the cranial segment of the esophagus. A tracheal cannula was inserted into the caudal portion of the esophagus where a tracheal fistula was made. Thereafter the patient continued to have respiratory problems but his respiratory condition improved gradually. The mode of mechanical ventilation was changed from intermittent mandatory ventilation (IMV) to continuous positive airway pressure (CPAP). However, he had repeated bronchitis, air trapping developed due to swelling around the tracheoesophageal fistula, and his lungs showed emphysema-like changes. The patient had cyanotic attacks and bradycardia, and expiratory rib cage compression was used to promote exhalation.

Subsequently, positive end expiratory pressure (PEEP) was slowly lowered. The patient was able to manage without the ventilator for progressively longer periods beginning at age 18 months. He initially needed PEEP because of the collapsing tracheoesophageal fistula. Other

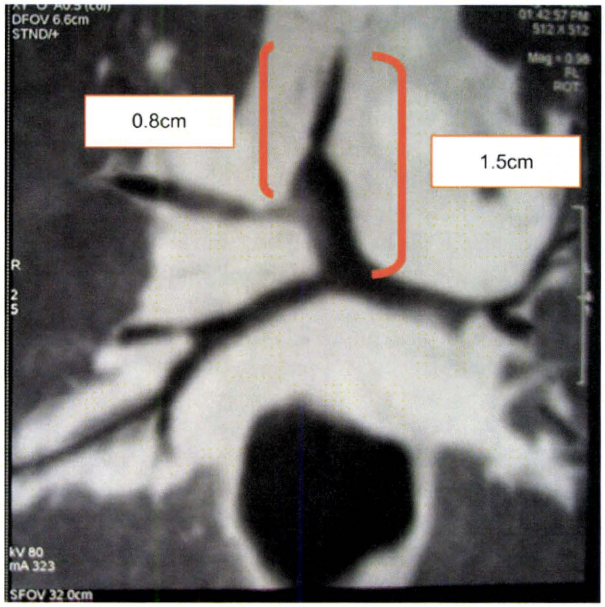

Fig. 2 3D-CT findings. Tracheal branching from the esophagus was narrowed at the superior portion. The tracheal bifurcation was 1.5 cm from the fistula. The right upper lobe bronchus arose in an area 0.8 cm from the fistula and formed a tracheal bronchus

procedures were also considered such as resection of stenosis of the fistula, end-to-end anastomosis, and placement of an external stent [4, 5]. The esophagus, tracheal fistula, and trachea were periodically examined by endoscopy. Occlusion of the fistula during straining and coughing gradually improved due to maturing tracheal cartilage and enlarging fistula diameter as the patient grew (Fig. 3b). The patient was in a stable respiratory condition and successfully being weaned from ventilator.

His respiratory condition was improving. Therefore, at age 3 years 2 months, the esophagus was reconstructed using a whole gastric tube by the retrosternal route to

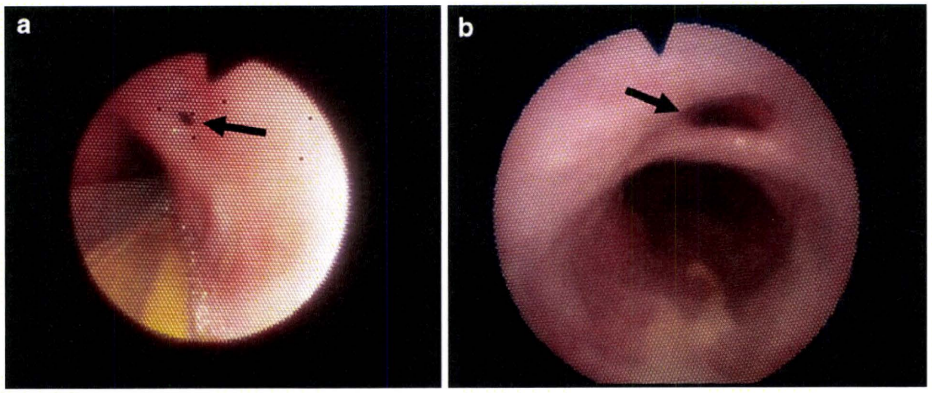

Fig. 3 Bronchoscopic findings. **a** At age 13 days, bronchoscopy revealed a tracheoesophageal fistula originating form the anterior esophageal wall, and the diameter of the fistula changed markedly with respiration. The image shows a very small diameter during expiration (*arrow*). **b** At age 2 years 10 months, the tissue had become sound around the opening of the tracheoesophageal fistula and changes in diameter were minimal during respiration (*arrow*)

enable oral feeding (Fig. 4a, b). Presently, the patient is receiving training in oral feeding. He has normal growth in height and weight and very good intellectual development. He uses the mechanical ventilator only at night, but he can manage without it most of the time.

Discussion

Tracheal agenesis was first reported by Payne et al. in 1900. Since then, over 140 cases have been reported in the literature. The majority of the cases died at birth or shortly thereafter. Most cases do not survive long term because of difficult airway management and serious associated disorders. In our search of the literature, we have found only four other long-term survival cases to date [2, 4–6]. Our patient had Floyd's type I tracheal agenesis, and airway management was difficult due to a fragile tracheoesophageal fistula. Conservative treatment was performed for the tracheoesophageal fistula, and the fistula became stable with the patient's physical growth. This patient is a very rare case in whom oral feeding was achieved after tracheal reconstruction using a gastric tube [6].

Floyd's classification of tracheal agenesis is the most commonly used system [3]. It divides the condition into three main types according to the form of the fistula, tracheoesophageal or bronchoesophageal fistula: type I (11% of all published tracheal agenesis cases), type II (61%), and

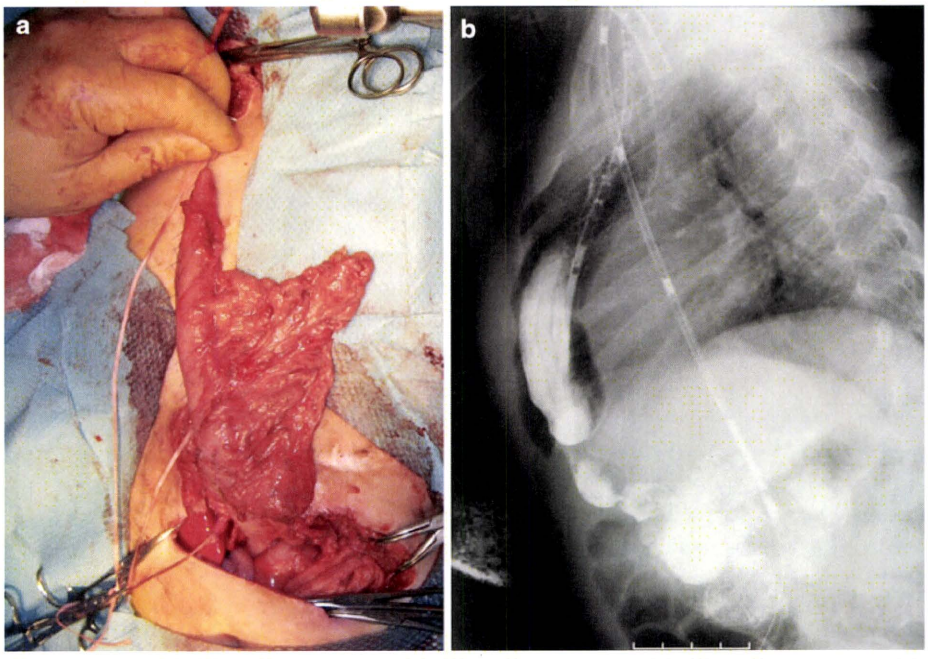

Fig. 4 **a** Esophageal reconstruction. A whole gastric tube was used for the esophageal reconstruction. Blood supply was from the gastroepiploic artery. **b** Oral contrast enhanced image taken after esophageal reconstruction

type III (23%) [7]. Types II and III have bronchi which directly formed a fistula with the esophagus, so the airway can easily occlude. In these types, tube insertion is more difficult into the tracheoesophageal or bronchoesophageal fistula and airway maintenance is also more difficult compared to type I. Most type I patients also die in the early neonatal period because of ventilatory failure due to esophageal collapse or accumulation of secretion.

In general, tracheal agenesis was suspected because of a history of antenatal polyhydramnios, absence of the first cry at birth, possibility of mask ventilation, and impossibility of endotracheal intubation, and was diagnosed by esophagography depicting the trachea, bronchoscopy showing tracheal atresia. If mask ventilation is possible and the infant can be successfully resuscitated immediately after birth, the next steps are to perform (1) abdominal esophageal banding for palliative surgery to substitute esophagus for trachea, (2) transection at the cervical esophagus with esophagostomy, and (3) gastrostomy for enteral feeding [8–11].

After palliative surgery is performed for substituting esophagus for trachea, the tracheoesophageal fistula should be treated in one of three major ways depending on the position and size of the fistula: (1) conservative management by follow-up, (2) direct insertion of the tip of an endotracheal tube into the fistula, or (3) surgical treatment of external stent placement after resection of the stenotic area and anastomosis. In the patient of the present report, intubation of the fistula was not selected because difficulties in its management were expected, including accidental extubation and sacrifice of the right tracheal bronchus. The tracheoesophageal fistula could easily collapse (Fig. 3a). In our case, ventilation improved with gradual mechanical stability of the tracheoesophageal fistula and with the patient's physical growth (Fig. 3b). Thus, his respiratory condition also stabilized.

Hiyama et al. placed an endotracheal tube in a fistula for a type II case and had successful respiratory management by CPAP ventilation. They performed esophageal reconstruction with colonic interposition between the residual esophagus and stomach [6]. We performed esophageal reconstruction on our patient at age 3 years 2 months to enable oral feeding. To our best knowledge, there have been only two cases, including our patient, who underwent esophageal reconstruction for tracheal agenesis. If small intestine or colon interposition for esophageal substitution is used in the reconstruction, there could be an ulcer on the intestinal side with a high risk for ischemia. Therefore, a gastric tube was used in our patient (Fig. 4a, b). In our route of reconstruction, intrathoracic anastomosis was not selected because respiratory failure was expected from swelling and collapse of the fistula due to surgical manipulation around it. Since a gastric tube was used, the suturing in the antethoracic route was thought to be impossible because of the distance. Therefore, the retrosternal route was selected. This route made it unnecessary to do any manipulation near the tracheoesophageal fistula during the surgery, and esophageal reconstruction was possible without thoracotomy. However, extracorporeal membrane oxygenation (ECMO) was set up so that it could be initiated any time an unexpected situation developed during the surgery.

Presently, our patient is being trained in oral feeding, and he has been gradually learning to swallow saliva. He had normal physical growth in height and weight due to enteral feeding and good intellectual and mental development. We await future developments in laryngotracheal transplant or regenerative medicine for treating his speech.

A written informed consent was obtained from the parents for submitting the manuscript. This study has been performed with the ethical standards laid down in the 1964 Declaration of Helsinki.

References

1. Hirakawa H, Ueno S, Yokoyama S, Soeda J, Tajima T, Mitomi T, Makuuchi H (2002) Tracheal agenesis: a case report. Tokai J Exp Clin Med 27:1–7
2. Soh H, Kawahawa H, Imura K, Yagi M, Yoneda A, Kubota A, Okada A (1999) Tracheal agenesis in a child who survived for 6 years. J Pediatr Surg 34:1541–1543
3. Floyd J, Campbell DC Jr, Dominy DE (1962) Agenesis of the trachea. Am Rev Respir Dis 86:557–560
4. Baroncini-Cornea S, Fae M, Gargiulo G, Gentili A, Lima M, Pigna A, Pilu G, Tancredi S, Turci G (2004) Tracheal agenesis: management of the first 10 months of life. Paediatr Anaesth 14:774–777
5. Watanabe T, Okuyama H, Kubota A, Kawahara H, Hasegawa T, Ueno T, Saka R, Morishita Y (2008) A case of tracheal agenesis surviving without mechanical ventilation after external esophageal stenting. J Pediatr Surg 43:1906–1908
6. Hiyama E, Yokoyama T, Ichikawa T, Matsuura Y (1994) Surgical management of tracheal agenesis. J Thorac Cardiovasc Surg 108:830–833
7. Lander TA, Schauer G, Bendel-Stenzel E, Sidman JD (2004) Tracheal agenesis in newborns. Laryngoscope 114:1633–1636
8. Fonkalsrud EW, Martelle RR, Maloney JV Jr (1963) Surgical treatment of tracheal agenesis. J Thorac Cardiovasc Surg 45:520–525
9. Altman RP, Randolph JG, Shearin RB (1972) Tracheal agenesis: recognition and management. J Pediatr Surg 7:112–118
10. Buchino JJ, Meagher DP Jr, Cox JA (1982) Tracheal agenesis: a clinical approach. J Pediatr Surg 17:132–137
11. Pratap A, Saha GS, Bhattarai BK, Yadav RP, Nepal A, Bajracharya A, Kumar A, Adhikary S (2007) Tracheal agenesis type B: further evidence to a lethal congenital tracheal malformation. J Pediatr Surg 42:1284–1287

PEDIATRICS INTERNATIONAL Official Journal of the Japan Pediatric Society
Pediatrics International (2011) **53**, 127–128 doi: 10.1111/j.1442-200X.2010.03283.x

Letter to the Editor

Survival of a congenital ileal atresia infant weighing 359 g at birth after laparotomy

Yasushi Fuchimoto,[1] Hirofumi Tomita,[1] Fumika Takasato,[1] Yuki Yamamoto,[1] Masaharu Mori,[1] Naoki Shimojima,[1] Ken Hoshino,[1] Isamu Hokuto,[2] Kazushige Ikeda[2] and Yasuhide Morikawa[1]

Departments of [1]Pediatric Surgery and [2]Pediatrics, School of Medicine, Keio University, Tokyo, Japan

The survival of extremely low-birthweight infants has dramatically improved with recent advancements in perinatal care. We report here a congenital ileal atresia infant weighing 359 g, considered to be the lowest birthweight among reported infant cases with laparotomy.

Because of threatened preterm labor, the infant was delivered via cesarean section at 26 weeks and 3 days of gestation with a birthweight of 359 g. Severe growth restriction had been found at the 19th gestational week. The fetus had stopped growing after the 24th gestational week at approximately 300 g. Fetal ultrasonography revealed a ventricular septal defect and single umbilical cord artery.

Tube feeding was initiated at 4 days of age, but the infant vomited and tube feeding was discontinued. Intestinal dilatation was found at 6 days of age, and dilatation gradually worsened. At the beginning, meconium-related ileus was suspected, and an amidotrizoic acid enema was administered but failed. Differential diagnosis included necrotizing enterocolitis, meconium related ileus, and intestinal atresia. It was difficult to finally diagnose ileal atresia until laparotomy was performed.

Thereafter gradual intestinal dilatation was observed and movement of gas stopped. When the patient was 10 days old, plain abdominal radiographs showed marked, localized intestinal dilatation (Fig. 1). There was a risk of intestinal perforation because of an intestinal obstruction. Therefore, emergency laparotomy was carried out after we confirmed respiratory and hemodynamic stability. We paid special attention to maintain blood pressure and urine output. During the operation, we made best efforts to prevent lowering of temperature. We performed these procedures in the operating room. An anesthesiologist who specialized in neonates administered the anesthesia.

In the laparotomy findings, marked intestinal dilatation was observed on the oral side of the atretic area. The distal end was approximately 20 cm proximal to the cecum. The proximal intestine was partially twisted and revealed necrosis, and the mesenteric defect was observed at this part. Therefore, we thought the cause of atresia was ischemic necrosis due to intestinal volvulus.

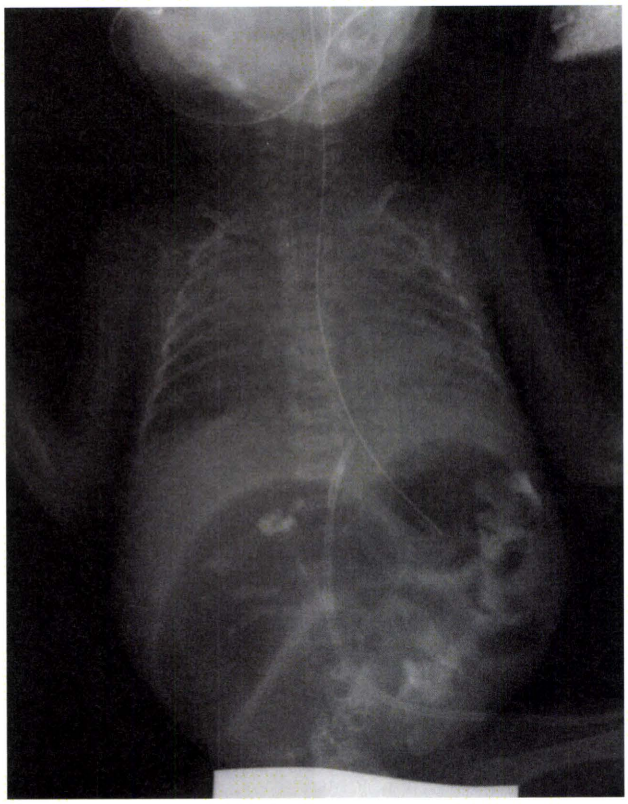

Fig. 1 Plain abdominal radiograph showed marked, localized intestinal dilatation at 10 days of age.

The necrotic area of the intestine was resected and ileostomy was performed. There was respiratory and hemodynamic stability during the surgery.

The infant was administered mother's milk after the operation, and fortified mother's milk was administered after achieving full feeding. After the infant's condition became stable, milk for low-birthweight children was added after the production of the mother's milk decreased.

Postoperative weight gain was satisfactory, and the postoperative course was uneventful without complications, such as wound infection or hemorrhage. The stoma was closed at 131 days of age (121 days postoperatively), when the infant was 1882 g in

Correspondence: Yasushi Fuchimoto, MD PhD, Department of Pediatric Surgery, School of Medicine, Keio University, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan. Email: yfuchimo@sc.itc.keio.ac.jp

Received 22 March 2010; revised 30 September 2010; accepted 4 October 2010.

weight. The milk was started on the 7th postoperative day, and oral feeding progressed smoothly thereafter.

Remarkable advancements in perinatal and neonatal care have enabled the survival of infants with extremely low birthweights, and the number of such infants is increasing. Presently, there is an increasing number of infants who can undergo surgical intervention because of advancements in neonatological management. These patients are infants who, in the past, would have died without an opportunity for surgical intervention. In surgical cases who survive, extremely low birthweight infants are reported to have poor neurological development compared to non-surgical cases.[1–3] Although prognosis was not favorable for neurological development of low birthweight infants, another report stated that the prognosis did not differ between infants with and without surgical intervention.[4]

In our case, evaluations are necessary not only for vital prognosis but also for neurological development over time. For such a case with an extremely low birthweight of approximately 350 g, survival is possible by surgery if the patient's general condition allows. This information is important in the process of informed decision-making.

To our best knowledge, this paper is a report of the lowest birthweight infant who underwent laparotomy.

References

1 Chacko J, Ford WD, Haslam R. Growth and neurodevelopmental outcome in extremely-low-birth-weight infants after laparotomy. *Pediatr. Surg. Int.* 1999; **15**: 496–9.
2 Hintz SR, Kendrick DE, Stoll BJ *et al.* Neurodevelopmental and growth outcomes of extremely low birth weight infants after necrotizing enterocolitis. *Pediatrics* 2005; **115**: 696–703.
3 Rees CM, Pierro A, Eaton S. Neurodevelopmental outcomes of neonates with medically and surgically treated necrotizing enterocolitis. *Arch. Dis. Child. Fetal Neonatal Ed.* 2007; **92**: F193–8.
4 Limpert JN, Limpert PA, Weber TR *et al.* The impact of surgery on infants born at extremely low birth weight. *J. Pediatr. Surg.* 2003; **38**: 924–7.