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Congenital H-type Tracheoesophageal Fistula:  
2 Cases Safely Diagnosed with Metrizamide

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Metrizamide を用いて安全に診断しえた食道閉鎖を  
伴わない気管食道瘻の2例

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食道閉鎖を伴わない気管食道瘻はきわめてまれな先天性疾患のひとつであるが、くりかえし認められる小児の肺炎においては必ず鑑別診断のひとつに加えねばならないものである。その診断はしかしながら容易ではなく、何回か造影剤を食道内に注入しなければならない場合が多い、そのような場合、造影剤は少なからず気管内、肺内へと吸引されその毒性が問題となる。今回我々は他院

にてイオン性ヨード剤が使用され、本院にてメトリザマイドを使用し確定診断しえた食道閉鎖を伴わない気管食道瘻を2例経験したので、それらをもとに文献的考察も加えて報告した。メトリザマイドは本疾患の診断にあたり、他の造影剤に比して安全に使用でき、また造影能にもすぐれたものであると考えられた。

### Introduction

Congenital H-type tracheoesophageal fistula (TEF), or congenital TEF without esophageal atresia is one of the less common developmental anomalies. A newborn infant with paroxysms of coughing or choking precipitated by feeding, abdominal distension, and recurrent pneumonia<sup>1)</sup> should raise the suspicion of a congenital H-type TEF. Radiographic evaluation with contrast agent to confirm a suspected congenital H-type TEF may result in aspiration of the contrast agent into the lung. Though no any contrast agent is completely innocuous, metrizamide causes fewer problems than other contrast agents and is better tolerated while aspirated into the lung<sup>2,3)</sup>. Two cases with suspected congenital H-type TEF are presented in whom metrizamide was used successfully for diagnosis without any complications.

### Case Reports

Case 1: A male patient was born on July 27, 1984 following a 40-week gestation and normal smooth

delivery. His birth weight was 2525 g and the apgar score was 9. On the next day, the patient was found not doing well, hypothermia, tachypnea and attacks of apnea developed. Neonatal infection was initially considered but the subsequent laboratory examinations showed lack of evidence of infection. On July 29 and 30, several spells of vomiting with cyanosis during feeding were noted. The chest roentgenogram revealed no obvious anomalies except for gaseous distension of the gastrointestinal tract (Fig. 1-A). An emergent esophagogram with hypertonic water-soluble contrast agent was performed by the surgeon. The contrast agent was found aspirated into the tracheobronchial trees, but it was uncertain whether the contrast agent was aspirated from the larynx or whether it passed through an H-type TEF. Respiratory distress appeared after the completion of the procedure, and the patient needed an endotracheal intubation and intensive treatment. The chest roentgenogram obtained at that time revealed bilateral ground glass appearance with prominent air bronchograms suggesting pulmonary edema (Fig. 1-B). A few days later, another esophagogram with metrizamide was done and successfully demonstrated a fistula at the level of the first thoracic vertebra (Fig. 2). The patient was uneventful after the examination. The chest roentgenogram showed no significant change comparing with that obtained before the esophagography (Fig. 3-A, B). On August 24, the operation through a cervical approach was performed and revealed an H-type TEF which was divided and closed. The postoperative course was smooth and a follow-up esophagogram showed no communication between the esophagus and tracheas.

Case 2: A male patient, a product of an uncomplicated, full-term gestation with birth weight of 2800 g, was born on June 5, 1984. He experienced choking and vomiting during the first feeding after delivery. A chest roentgenogram obtained on the next day revealed infiltrates of both upper lobes and right middle lobe of the lung (Fig. 4). Because choking and vomiting were associated with the further oral feedings, an

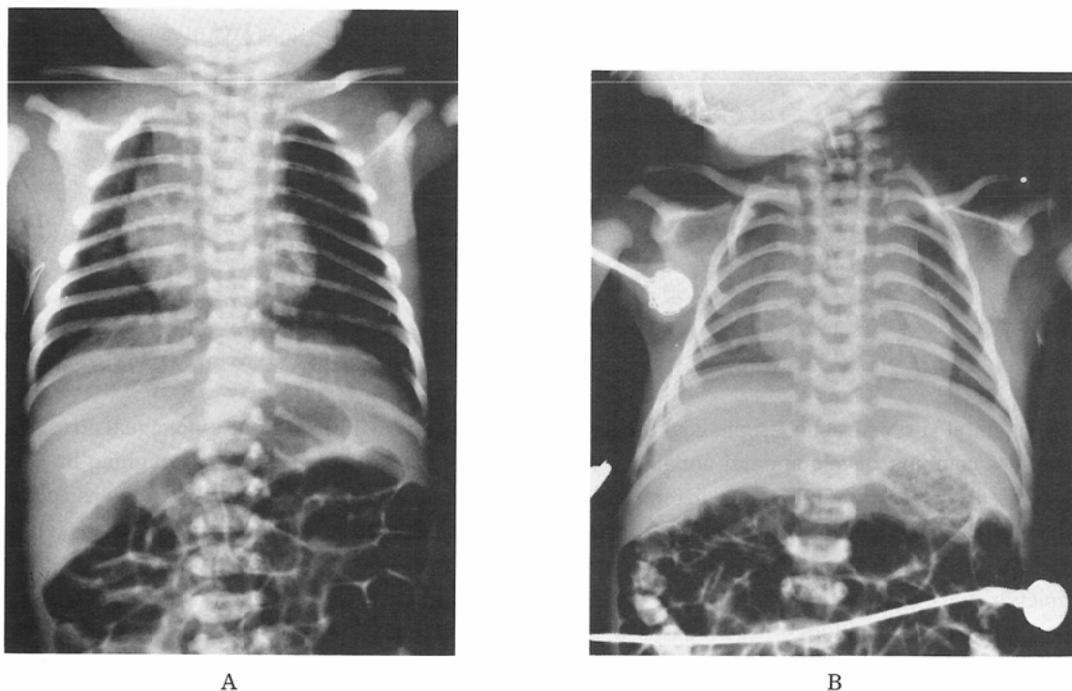


Fig. 1 Case 1. Chest roentgenogram. A, Before esophagogram with Urografin, note gaseous distension of the GI tract. The lung field is clear. B, After the esophagogram with Urografin, note that there is bilateral ground glass appearance with prominent air bronchograms, representing pulmonary edema. Also note an endotracheal tube in the trachea.

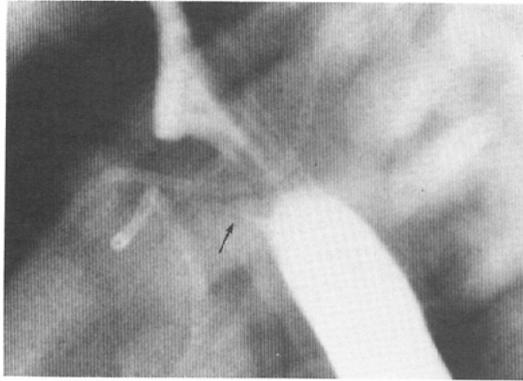
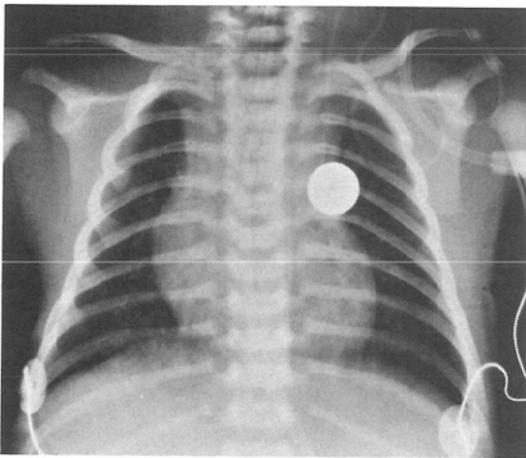
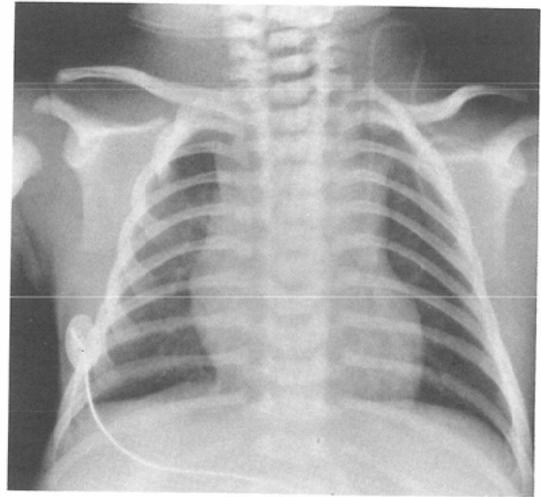


Fig. 2 Case 1. Esophagogram with metrizamide demonstrates an H-type TEF at the level of the first thoracic vertebra (arrow). Note initial filling of esophageal end of the fistula.



A



B

Fig. 3 Case 1. Chest roentgenogram. A, Before esophagogram with metrizamide shows clear lung field. B, After esophagogram with metrizamide, note that there is no evidence of pulmonary edema or residual contrast media.

esophagogram with Urografin was performed on June 21. Urografin was seen to enter the tracheobronchial trees and was considered as aspiration. After controlling the aspiration pneumonia with antibiotics and nasopharyngeal tube feeding, the patient was transferred to Keio University Hospital for further evaluation on August 22. Shortly after admission, an esophagogram using metrizamide was done which successfully showed a fistula at the level of thoracic inlet running anteriorly and upward to enter the trachea (Fig. 5-A, B). The chest roentgenogram obtained after the procedure showed no significant change (Fig. 6-A, B). On September 5, the patient underwent surgical repair through a cervical approach. The fistula was visualized, divided and closed. A postoperative esophagogram also performed and showed no evidence of leak or recurrence of the fistula.

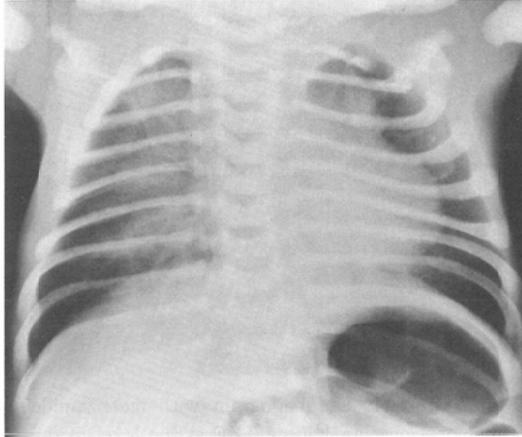


Fig. 4 Case 2. Chest roentgenogram shows aspiration pneumonia. There are widespread infiltrates of both upper lobes and right middle lobe.

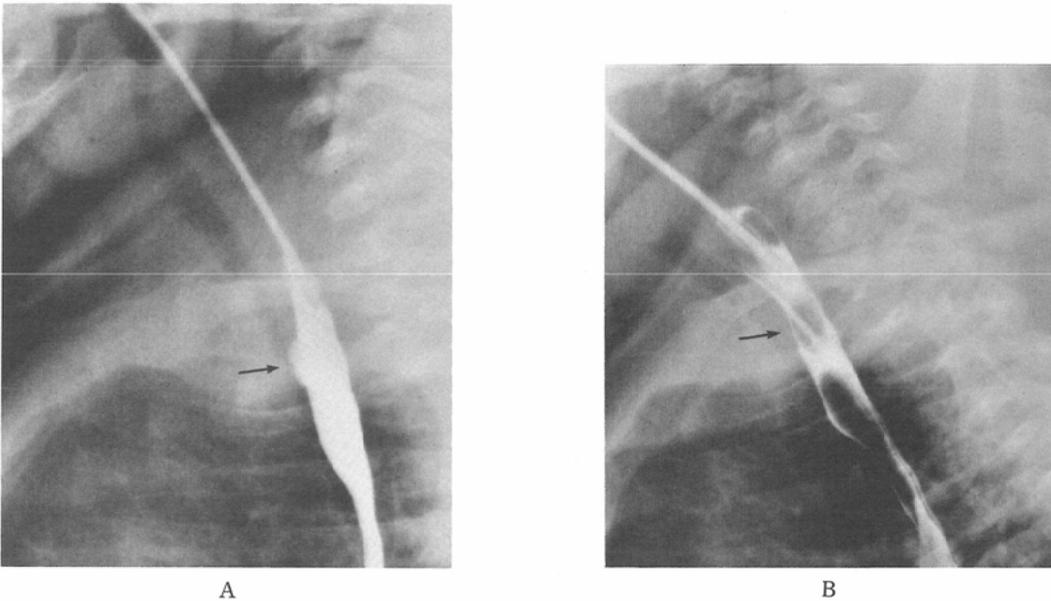


Fig. 5 Case 2. Esophagogram with metrizamide. A, Note that there is an outpouching of the esophagus at the origin of the fistula (arrow). B, Note an H-type TEF at the level of the first thoracic inlet (arrow).

### Discussion

Congenital H-type TEF, comprising about 4–6%<sup>4)5)</sup> of all congenital TEF, occurs in approximately one in 150,000 births<sup>6)</sup>. The rarity of this lesion is illustrated by the incidence of only one case every 12 to 18 months at referral hospitals<sup>7)8)</sup>. The first case of congenital H-type TEF which was diagnosed at autopsy in a 7-week-old infant was described by Lamb<sup>9)</sup> in 1873. Imperatori in 1939 was the first to make an exact preoperative diagnosis and to cure the patient by operation<sup>10)</sup>.

The precise etiology of the congenital H-type TEF is not completely understood. One theory explaining

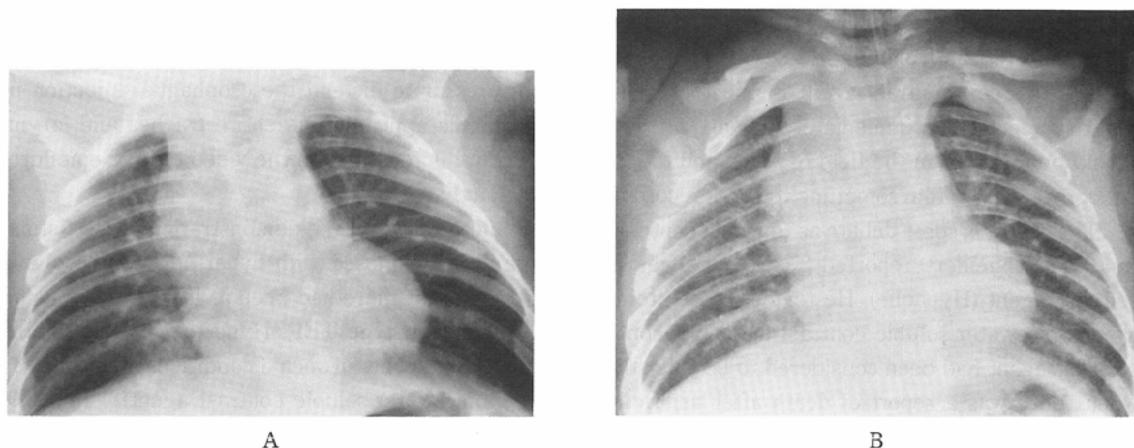


Fig. 6 Case 2. Chest roentgenogram. A, Before esophagogram with metrizamide reveals some pneumonic infiltration in both lung fields. B, After esophagogram with metrizamide, note that there is no evidence of pulmonary edema or residual contrast media.

the embryologic development of tracheoesophageal fistula hypothesizes that it is due to developmental error in formation and separation of primitive gut into trachea and esophagus. This developmental error, which may result from genetic<sup>11)</sup>, infectious, nutritional, toxic, metabolic, or physical influences<sup>12)</sup>, occurs probably during the third to sixth week of fetal life. There may be coexisting anomalies occurring at the same time of the intrauterine life with the incidence of 23%<sup>5)</sup> of congenital H-type TEF. Embryologically and clinically, the congenital H-type TEF represents the least severe degree of laryngotracheoesophageal cleft<sup>13)</sup>. The fistulous communication is most often in the upper and middle trachea. Johnston and Hastings<sup>14)</sup> reported that the level of the fistula was located at or above the secondary thoracic vertebra in 83% of total 63 cases reviewed from the literature and their own cases. Andrassy et al<sup>7)</sup> found that 70% fistulas were within 2 cm above or below the sternal notch, 17% were more than 2 cm above, and only 13% were more than 2 cm below the sternal notch.

The clinical picture of the congenital H-type TEF is depend on the size of the opening of the fistula. If the opening of the fistula is small, the clinical picture may be not so striking, and some patients may remain undiagnosed even into the late childhood. On the other hand, if the opening is large enough, clinical picture is severe and presents early. Helmworth and Pryles<sup>1)</sup> emphasized the triad of symptoms and signs including 1) paroxysms of coughing precipitated by feeding, 2) gaseous distension of the gastrointestinal tract, and 3) pneumonitis. Paroxysms of coughing precipitated by feeding is the most common symptom. Andrassy<sup>7)</sup> reported that it appeared in 90% of their cases. It should be noticed that in some patients of severely ill, low birth weight, or premature, cough reflex may be weak or even absent, and the presenting symptom would be vomiting or cyanosis during feeding rather than coughing. The next common symptom is aspiration pneumonia, occurring in 81% of cases<sup>7)</sup>. Roentgenographically, the pattern of aspiration pneumonia in congenital H-type TEF usually shows widespread infiltrates, and right upper lobe aspiration is more common in esophagela atresia<sup>15)</sup>. Gaseous distension of the gastrointestinal tract because of air entering the esophagus through the fistula was noted in 38% of patients<sup>7)</sup>. Excessive nasopharyngeal mucus was the another symptom reported by Andrassy<sup>7)</sup> and Bedard<sup>8)</sup> in 62% and 39% of cases respectively.

Congenital H-type TEF can be suspected on the basis of rather definite clinical symptoms and signs but may be difficult to prove by roentgenographic evidence, and is a challenge to the radiologist. The esophagogram with contrast agent is usually taken first in an attempt to confirm the diagnosis.

Congenital H-type TEF is demonstrated with the patient in right lateral decubitus position under

fluoroscopy. An 8 French nasogastric tube is placed through the nose to the distal esophagus. The tip of the tube is pulled back from the distal esophagus to proximal esophagus slowly. During pulling back the tube, at every 1 to 2 cm, contrast agent is injected with moderate pressure to distend the esophagus. Injection of contrast agent stops immediately when the trachea or the fistula appears. The entire trachea and larynx should be included in the field of view, and the whole procedure is recorded on the video tape or cinefluorography. Spot film sometimes can not record the fistula exactly.

There still is some debate as to which contrast agent should be used for the demonstration of the fistula. In 1961, Schneider<sup>6)</sup> reported 2 cases of congenital H-type TEF diagnosed with hypertonic water-soluble contrast agent (Hypaque). He reviewed the literature and found that there had been at least 3 cases using hypertonic water-soluble contrast agent for confirming congenital H-type TEF. Hypertonic water-soluble contrast agent had been considered to be safer than other contrast agents in such a condition<sup>19)20)</sup> until 1968 when there was a report of death after aspiration of hypertonic water-soluble contrast agent<sup>21)</sup>. In 1969, Reich<sup>22)</sup> investigated an animal study and found that hypertonic water-soluble contrast agent was dangerous while it was aspirated into the lung because it might produce pulmonary edema. In 1978, the American Academy of Pediatrics Committee on Radiology<sup>23)</sup> suggested that hypertonic water-soluble contrast agent should be avoided when studying the upper gastrointestinal tract if there was any risk of pulmonary aspiration. In the lung, hypertonic water-soluble contrast agent causes severe pulmonary edema and pneumonitis. It is the main pathogenesis of respiratory distress and hypoxia while hypertonic water-soluble contrast agent is aspirated into the lung<sup>23)</sup>.

Barium sulfate suspension has been shown to be a relatively safe contrast agent, while aspirated into the tracheobronchial tree in small amount<sup>16)17)18)</sup>. However, if a large amount of barium sulfate, or if the infant with compromised lung function (not uncommon in the patient with congenital H-type TEF), pulmonary aspiration may enhance the morbidity or even result in fatal consequence. Instillation of barium sulfate suspension into the lung of rats and dogs was reported<sup>9)</sup> to produce profound alteration in blood gas, segmental atelectasis, focal pneumonitis and fibrosis. Barium sulfate was only slowly cleared by macrophages. This slow clearance of barium sulfate in the lung may also cause difficulty in the interpretation of the subsequent chest roentgenograms.

An ideal bronchographic contrast agent is not always an ideal contrast agent to demonstrate the tracheoesophageal fistula. Contrast agent used for bronchography usually has high viscosity, therefore it is difficult to pass through the small fistula and result in a negative examination.

Metrizamide which had been widely used for myelography, was first used as a diagnostic contrast agent in the gastrointestinal tract by Cohen<sup>24)</sup> in 1980. Later, some authors<sup>25)~27)</sup> also had a preference for using metrizamide in the condition of potential entry of contrast agent into the lung or potential leak of contrast agent from gastrointestinal tract. Metrizamide is a non-ionic, water-soluble iodine-containing contrast agent which can be used in isotonic solution. Isotonic metrizamide does not cause harmful fluid shift while aspirated into the lung. In animal study<sup>23)</sup>, metrizamide has shown to produce less histologic reaction, hypoxia or respiratory distress than barium sulfate and hypertonic water-soluble contrast agent. The clearance of metrizamide in the lung also is rapid and faster than barium sulfate and hypertonic water-soluble contrast agent. In comparison with other contrast agent, metrizamide is rather innocuous to the lung and is the contrast agent of choice in condition which the possibility of pulmonary aspiration exists.

Metrizamide with concentration of 180 mg I/ml was reported to have good visualization in the examination of esophagus, stomach, and intestine<sup>24)~27)</sup>. However, the resolution of the fluoroscopic monitor is not good enough to provide satisfactory visualization of metrizamide with concentration of 180 mg I/ml, especially in a small fistula. Spot radiography can provide good visualization of a small fistula even the concentration of metrizamide is lower than 180 mg I/ml. However, the timing of spot radiography may fail to detect the fistula, if the filling and emptying of the fistula by the contrast agent is almost instantaneously.

Cinefluroradiography can record the fistula in such condition with satisfactory visualization. If the cinefluroradiography is not available, higher concentration of the metrizamide should be used to provide good visualization in the fluoroscopic monitor. In our experience, metrizamide with concentration of 250 mg l/ml, though it is slightly hypertonic, is still rather safe to the lung, and can be visualized satisfactorily in the fluoroscopic monitor.

Surgical repair is the only method to cure the disease. About 87% of cases could be repaired through a cervical approach<sup>7)</sup>. Preoperative respiratory problems such as aspiration, pneumonia atelectasis and using inappropriate diagnostic contrast agent usually account for the early postoperative deaths. The risk now can be minimized by early diagnosis with metrizamide.

### Conclusion

1) Contentital H-type TEF is an uncommon developmental anomaly. 2 cases of congenital H-type TEF diagnosed with metrizamide are reported.

2) In view of the safety and usefulness, metrizamide has many advantages over other contrast agents, and is the contrast agent of choice to confirm a suspected congenital H-type TEF.

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