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ORIGINAL ARTICLE

CT findings of plastic bronchitis in children after a Fontan operation

Hyun Woo Goo · Won Kyoung Jhang · Young Hwee Kim · Jae Kon Ko · In Sook Park · Jeong-Jun Park · Tae-Jin Yun · Dong-Man Seo

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Abstract
Background Plastic bronchitis is a rare cause of acute obstructive respiratory failure in children. Life-threatening events are much more frequent in patients with repaired cyanotic congenital heart disease, and most frequent following a Fontan operation. Commonly, the diagnosis is not made until bronchial casts are expectorated. Detailed CT findings in plastic bronchitis have not been described.
Objective To describe the CT findings in plastic bronchitis in children after a Fontan operation.
Materials and methods Three children with plastic bronchitis after a Fontan operation were evaluated by chest CT. Bronchial casts were spontaneously expectorated and/or extracted by bronchoscopy. Airway and lung abnormalities seen on CT were analyzed in the three children.
Results CT demonstrated bronchial casts in the central airways with associated atelectasis and consolidation in all children. The affected airways were completely or partially obstructed by the bronchial casts without associated bronchiectasis. The airway and lung abnormalities rapidly improved after removal of the bronchial casts.
Conclusion CT can identify airway and lung abnormalities in children with plastic bronchitis after a Fontan operation. In addition, CT can be used to guide bronchoscopy and to monitor treatment responses, and thereby may improve clinical outcomes.

Keywords Plastic bronchitis · CT · Congenital heart disease · Surgery · Fontan operation · Children

H. W. Goo (✉)
Department of Radiology and Research Institute of Radiology,
Asan Medical Center, University of Ulsan College of Medicine,
388-1 Poongnap-2 dong, Songpa-gu,
Seoul 138-736, South Korea
e-mail: hwgoo@amc.seoul.kr

W. K. Jhang · Y. H. Kim · J. K. Ko · I. S. Park
Department of Pediatric Cardiology, Asan Medical Center,
University of Ulsan College of Medicine,
Seoul, South Korea

J.-J. Park · T.-J. Yun · D.-M. Seo
Department of Pediatric Cardiac Surgery, Asan Medical Center,
University of Ulsan College of Medicine,
Seoul, South Korea

Introduction

Plastic bronchitis is a rare condition characterized by bronchial casts in the central airways that may result in acute respiratory failure. Plastic bronchitis usually occurs in children with various underlying diseases, including respiratory infection, asthma, cystic fibrosis, sickle cell disease, cyanotic congenital heart disease and lymphatic abnormalities [1]. Of note, life-threatening events are much more frequent in patients with repaired cyanotic congenital heart disease, and most frequent following a Fontan operation [2]. Because clinical and radiological findings are nonspecific, the diagnosis of plastic bronchitis is usually delayed and usually made when bronchial casts are spontaneously expectorated or removed by bronchoscopy. To the best of our knowledge, detailed CT findings in plastic bronchitis have not been described. We describe here the CT findings in plastic bronchitis in three children after a Fontan operation.

Materials and methods

Three children with plastic bronchitis after a Fontan operation with an extracardiac conduit were evaluated by contrast-enhanced chest CT using 16-slice scanners.

Table 1 Demographics, underlying cardiac defects, and morphological features of bronchial casts in three children with plastic bronchitis after a Fontan operation.

Patient	Sex	Age at CT (years)	Interval between Fontan operation and CT (months)	Cardiac defects	Bronchial casts		
					Number	Location	Pattern of airway obstruction
1	F	4	13	Transposition of the great arteries, ventricular septal defect, pulmonary stenosis, criss-cross heart	Two	Right upper lobe and right lower lobe bronchi	Partial
2	F	4	13	Right isomerism, dextrocardia, complete atrioventricular septal defect, pulmonary atresia	One	Left main bronchus	Complete
3	M	4	12	Right isomerism, total anomalous pulmonary venous return, complete atrioventricular septal defect, pulmonary stenosis, criss-cross heart	Two	Basal segmental bronchi of both lower lobes	Partial

They presented with acute respiratory difficulty and cyanosis. Demographics and underlying cardiac defects in the three children are shown in Table 1. All three children suffered from prolonged chylothorax after a Fontan operation for 19 days, 38 days and 34 days, respectively. Body weight-based CT parameters were used as follows: 80 kVp, 65–90 effective mAs; 375-ms gantry rotation time, 1.5-mm detector collimation with 0.7-mm reconstruction interval, and pitch 1.0. Iodinated contrast agent was injected intravenously using both arm and leg veins at the same time to evaluate the patency of the Fontan pathway. Detailed CT techniques for congenital heart disease have been described previously [3, 4]. Because of their young age and severe respiratory difficulty, the children could not hold their breath during scanning. The diagnosis of plastic bronchitis was made when bronchial casts were spontaneously expectorated and/or extracted by bronchoscopy. In addition to the Fontan pathway, airway and lung abnormalities seen on CT were analyzed in the three children.

Results

In all three children the Fontan pathway was seen to be patent on CT (Fig. 1) and cardiac function was good on echocardiography. CT demonstrated bronchial casts in the central airways with associated atelectasis and consolidation in all children (Fig. 2). The locations of the bronchial casts varied from major bronchi to segmental bronchi (Table 1; Fig. 2). The affected airways were completely or partially obstructed by the bronchial casts without associated bronchiectasis. The airway and lung abnormalities rapidly improved after removal of the bronchial casts by either spontaneous

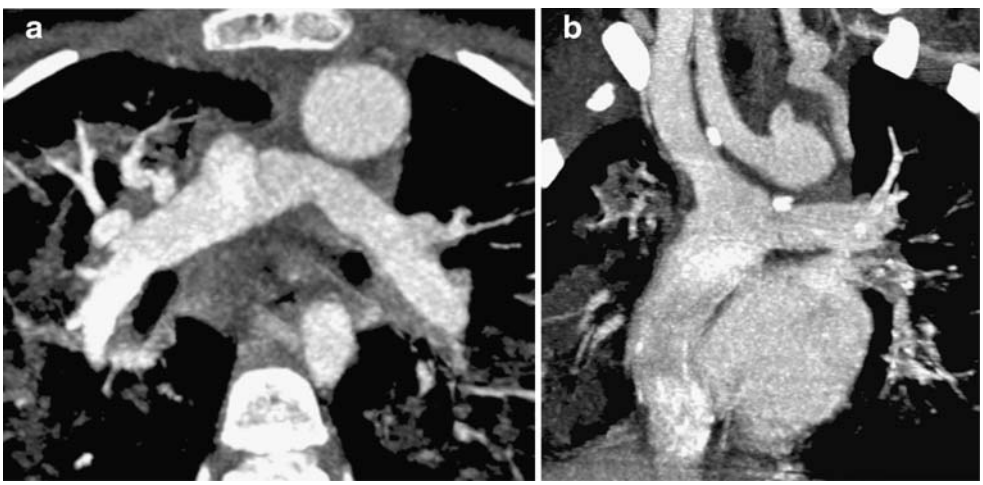

Fig. 1 Patency of the Fontan operation. Oblique axial (**a**) and oblique coronal (**b**) contrast-enhanced CT images show the patent Fontan pathway including the superior vena cava, the pulmonary arteries, and extracardiac conduit in a child with plastic bronchitis (patient 1). Areas of bilateral peribronchial consolidation are apparent

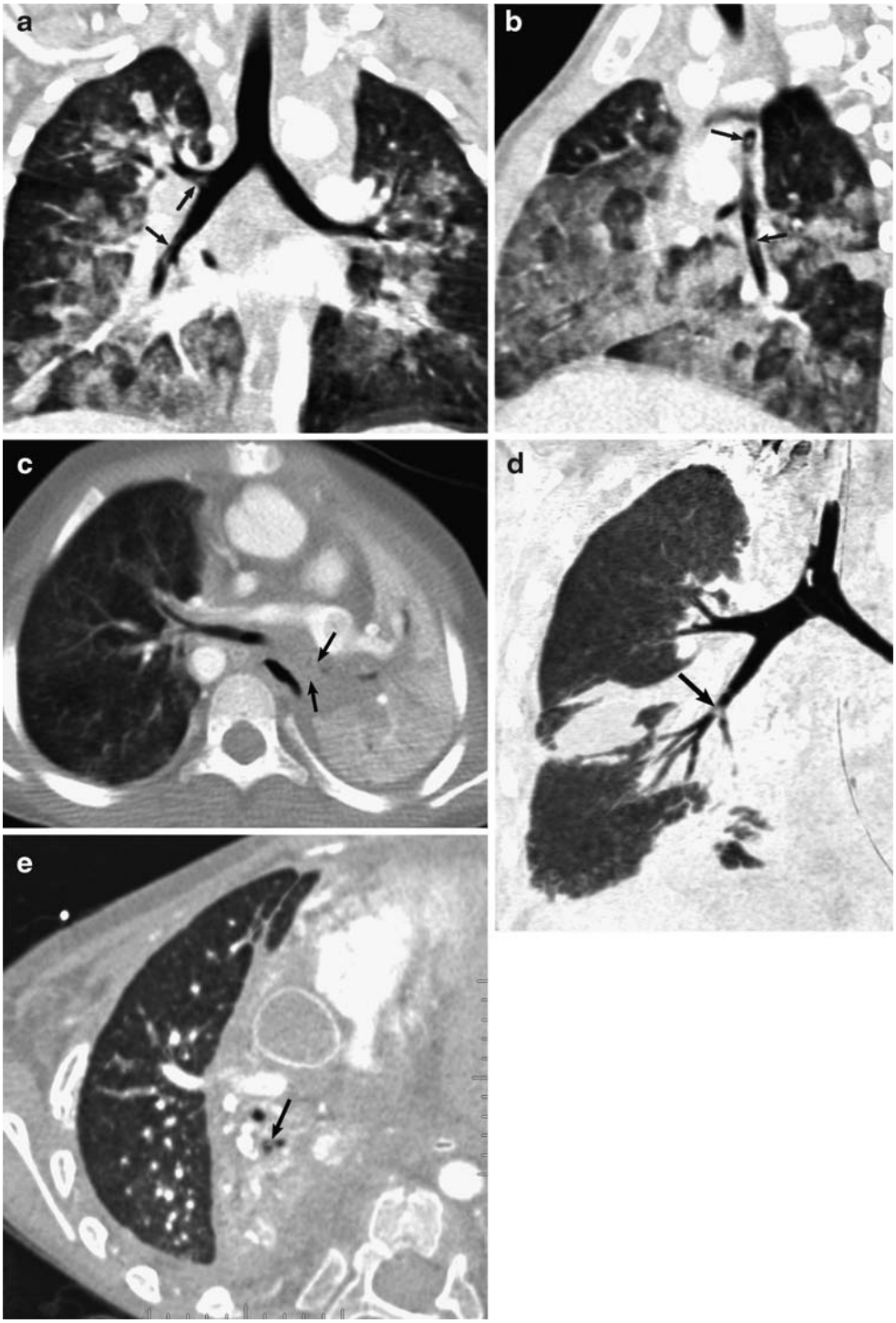

Fig. 2 CT appearances of bronchial casts. **a**, **b** Coronal (**a**) and sagittal (**b**) CT images with lung window setting reveal two bronchial casts (*arrows*) in a child with plastic bronchitis (patient 1), one in the right upper lobe bronchus and the other in the right lower lobe bronchus. Multiple bilateral areas of patchy consolidation or ground glass opacity are apparent. **c** Axial CT image in a different child with plastic bronchitis (patient 2) shows complete obstruction of the left main bronchus by a large bronchial cast (*arrows*) with total collapse of the left lung. The right descending thoracic aorta is seen. **d**, **e** Oblique coronal (**d**) and oblique axial (**e**) CT images with lung window setting demonstrate residual small bronchial casts (*arrow*) in the posterior basal segment of the right lower lobe after an episode of self expectoration in a child with plastic bronchitis (patient 3)

expectoration or bronchoscopic extraction (Fig. 3). CT was useful to guide bronchoscopy. The bronchial casts were whitish yellow and tenacious with a rubber-like consistency and they looked like the bronchial tree from which they had arisen. Histologically, fibrin and inflammatory cells were the main components of the casts (Fig. 4).

Rapid clearance of the radiological abnormalities was accompanied by a similar improvement in the clinical findings during treatment that included chest physiotherapy, acetylcysteine nebulizer, macrolide antibiotics, bronchoscopic extraction, urokinase nebulizer and subcutaneous heparin injection. Two of the three children experienced minor recurrent episodes of plastic bronchitis with similar presentations to the first episode after variable intervals. Awareness of plastic bronchitis allowed us to make the diagnosis of recurrent plastic bronchitis earlier than during

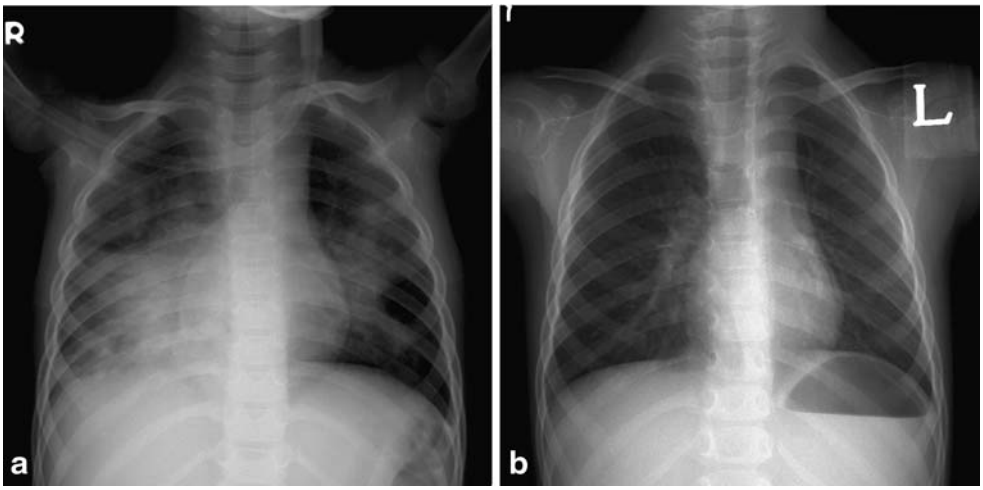

Fig. 3 Improvement after expectoration in a child with plastic bronchitis after a Fontan operation (patient 1). **a** Initial chest radiograph shows bilateral parahilar consolidation. Consolidation in the right lung is more extensive than in the left lung. **b** Follow-up chest radiograph after spontaneous expectoration of bronchial casts and medical treatment demonstrates complete resolution of previous lung lesions

the first episode, which might explain the milder condition. Treatment of the recurrent episodes was based on the relative effectiveness of previous treatments.

Discussion

CT was able to demonstrate bronchial casts in three children with plastic bronchitis after a Fontan operation. The bronchial casts identified in the central airways on CT might explain the obstructive nature of acute respiratory difficulty in our patients. This obstructive nature of the acute respiratory failure may mimic foreign body aspiration in children [5]. Lung parenchymal abnormalities were present in the affected lung regions. In contrast to mucoid impaction or other chronic airway infection, bronchiectasis was absent in our patients. Such detailed CT findings of plastic bronchitis, to the best of our knowledge, have not previously been reported. Chest radiographic and bronchographic findings have been reported to be nonspecific. These findings include collapse of the affected segment, obstructive hyperinflation, bilateral patchy consolidations, abrupt termination of a bronchus, bronchiectasis, pleural effusion, and air leaks [6, 7]. A "finger-in-glove" pattern reflecting widespread plugging of central airways was the only CT finding of plastic bronchitis previously described in the literature [1].

Mucoid impaction and foreign body aspiration are the main differential diagnoses of plastic bronchitis. The bronchial casts in plastic bronchitis tend to be larger and more cohesive than ordinary mucus plugs [8]. In contrast to plastic bronchitis, mucoid impaction is likely to be retained in airways rather than expectorated, and neither large nor branching in cases of expectoration [6]. Rapid clinical improvement after expectoration or bronchoscopic removal of bronchial casts and little evidence of ongoing bronchoconstriction may be clues to plastic bronchitis. In foreign body aspiration, a relevant clinical history is usually present or highly suggestive of the diagnosis. Rarely, a bronchial tumor cast has been reported in a child [9].

Seear et al. [1] classified plastic bronchitis into two types: type I (inflammatory cast consisting of fibrin, eosinophilic inflammatory infiltrates, and Charcot-Leiden crystals), developed in infection, asthma, cystic fibrosis, and sickle cell disease; and type II (acellular cast consisting

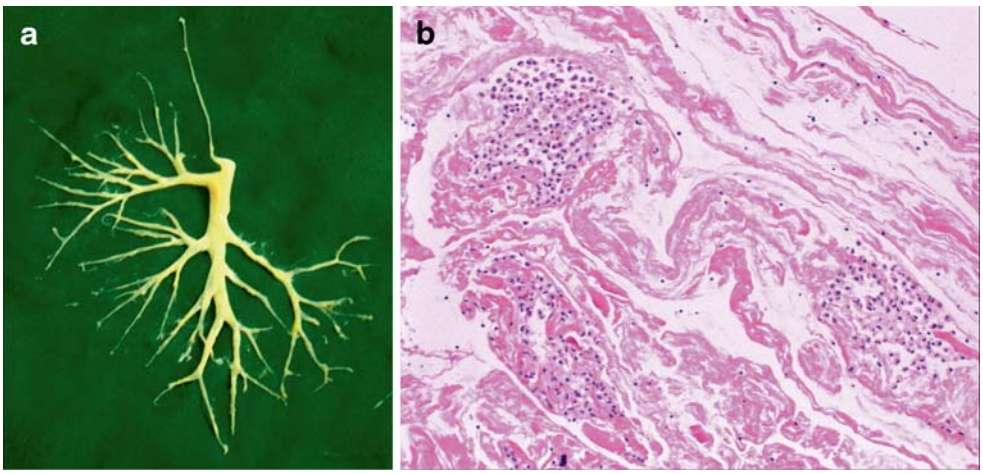

Fig. 4 A spontaneously expectorated bronchial cast from a child with plastic bronchitis after a Fontan operation (patient 1). **a** The cast looks like a whitish yellow bronchial tree. **b** Histologically, the cast is mainly comprised of fibrin and inflammatory cells (H&E, ×100)

mainly of mucin), developed in cyanotic congenital heart diseases (typically after surgical repair, the most frequent being a Fontan operation, followed by Blalock-Taussig shunt) and lymphatic abnormalities. The proposed pathogenesis of type II casts includes hypersecretion of airway mucus [1], abnormalities of pulmonary lymphatic drainage resulting in lymphatic leakage into the bronchi [10], and poor cardiac output.

A new classification scheme was recently proposed based on the associated disease and cast histology when comorbidity is unclear [11]. The classification scheme includes four categories: (1) structural congenital heart diseases and mucin predominant casts; (2) lymphatic disorders and chylous casts; (3) asthma, atopy and eosinophilic casts; and (4) acute chest syndrome in sickle cell disease and fibrinous casts. Despite the bronchial casts in our patients mainly consisting of fibrin and inflammatory cells, the casts belonged to the first category according to the new classification of plastic bronchitis because our patients had structural congenital heart disease and had undergone a Fontan operation.

The Fontan pathway and cardiac function should be evaluated in a patient with suspected plastic bronchitis after a Fontan operation because hemodynamic disturbance is one of the pathogenetic mechanisms in plastic bronchitis. Improvement in hemodynamic disturbance may improve the disease condition. Hemodynamic disturbance as a cause of plastic bronchitis was excluded in our patients by contrast-enhanced CT and echocardiography. Prolonged postoperative chylothorax remained a potential cause of the plastic bronchitis in our patients. Although time-resolved, three-dimensional contrast-enhanced MR angiography using dilute contrast agent is the procedure of choice for evaluating patency and flow dynamics of the Fontan pathway [12], CT is definitely preferred to MR angiography when plastic bronchitis is suspected after a Fontan operation, as in our patients. According to a review of 42 reported patients [2], congenital heart disease is the most common underlying disease (40%). In addition, life-threatening events are substantially more frequent in patients with congenital heart disease (41%) than in those with asthma (0%) [2].

We speculate that the true incidence of plastic bronchitis is likely to be underestimated because of difficulty in its recognition. Therefore, CT should be performed promptly in children showing acute respiratory failure after a Fontan operation to avoid a delay in the diagnosis of plastic bronchitis, a treatable disease. CT is also useful to guide bronchoscopy, a diagnostic and therapeutic procedure for plastic bronchitis. Therapeutic options for plastic bronchitis are diverse and no one therapy is effective; therapeutic responses are quite different among individuals with plastic bronchitis. Therefore, a vigorous search for an effective therapeutic modality is necessary and should be individualized.

Conclusion

CT can identify airway and lung abnormalities in children with plastic bronchitis after a Fontan operation. In addition, CT can be used to guide bronchoscopy and to monitor treatment response, and thereby may improve clinical outcome in patients with plastic bronchitis.

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