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Resolution of Plastic Bronchitis with Atrial Pacing in a Patient with Fontan Physiology

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Abstract. We describe a 5-year-old boy with Fontan physiology and a ventricular pacemaker who developed severe plastic bronchitis. Evaluation by cardiac catheterization revealed profoundly altered hemodynamics, which improved with atrial pacing. Following implantation of an atrial pacemaker, which restored atrioventricular (A-V) synchrony, the patient's hemodynamics greatly improved and his plastic bronchitis resolved.

Keywords: Plastic bronchitis — Atrial pacing — Fontan physiology

First described in 1902 [1], plastic bronchitis is a rare, life-threatening condition in which large, arboreal bronchial casts obstruct the airway. Histologically the casts contain eosinophils or, when associated with congenital heart disease, may be acellular [10]. Plastic bronchitis has been described with a variety of disorders including patients with Fontan circulation. Multiple treatments have been proposed to treat this condition, including antibiotics, steroids, bronchodilators, mucolytic agents, anticoagulation, aerosolized urokinase, and tissue plasminogen activator, with anecdotal success.

The optimal treatment for this condition remains unclear, but medications and interventions associated with improved hemodynamics have led to resolution of this disorder [3, 8, 9, 11]. The optimal mode for pacing patients with Fontan circulation has not been formally studied. Our case demonstrates the importance of A-V synchrony in Fontan patients with altered hemodynamics.

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Case Report

A 5-year-old boy with tricuspid atresia, who underwent a lateral tunnel, fenestrated Fontan procedure at age 4, was referred to our center secondary to complications of plastic bronchitis. Following his Fontan procedure he developed a slow junctional rhythm, and 1 month after his surgery, a ventricular (VVI) epicardial pacemaker was implanted. He subsequently developed increasing respiratory symptoms and 6 months after his Fontan his cough became productive for mucinous bronchial casts (Fig. 1). The patient was hospitalized several times for pneumonias and underwent several bronchoscopies with bronchial cast aspiration and also developed recurrent pericardial effusions requiring multiple pericardiocenteses.

At the time of referral to our center he had been hospitalized for the majority of the previous several months and was experiencing daily bouts of severe coughing fits, with profound desaturation and bronchial cast expectoration. His examination revealed an ill-appearing, cyanosed young boy in moderate respiratory distress. Oxygen saturations on 2 liters of oxygen were in the 70s and dropped to the 50s with bouts of coughing. Lungs revealed pronounced bilateral crackles to auscultation, and his chest x-ray was markedly abnormal (Fig. 2). The patient underwent cardiac catheterization for hemodynamic assessment and evaluation of the effects of atrial versus ventricular pacing. During the catheterization, ventilation and oxygenation proved increasingly difficult secondary to airway obstruction by bronchial casts. Despite endotracheal intubation and frequent suctioning, emergent bronchoscopy with prolonged, deep suctioning, performed on multiple occasions, was required to clear the airway. Baseline hemodynamics with VVI pacing at 110 beats/min (Fig. 3A) revealed markedly elevated left atrial pressures with cannon waves (peak, 22; mean, 8 mmHg) and a pulmonary artery pressure of approximately 18 mmHg with a 2-Torr gradient from the SVC to the right pulmonary artery. With atrial (AAI) pacing at 110 beats/min, associated with intact A-V conduction and restoration of atrioventricular synchrony, hemodynamics markedly improved (Fig. 3B). We noted immediate dissolution of the left atrial cannon waves, restoration of normal left atrial A and V waves (A, 5; V, 5; mean, 4 mmHg), mild reduction of the pulmonary artery pressure to 16 mmHg, and mild reduction of left ventricular end diastolic pressure (6 mmHg with VVI to 4 mmHg in AAI mode). Systemic blood flow also increased by 50% with institution of atrial pacing.

After hemodynamic assessment, a stent was placed across the right pulmonary artery and the 2-Torr gradient was relieved. The patient had an atrial pacemaker implanted and gradually improved

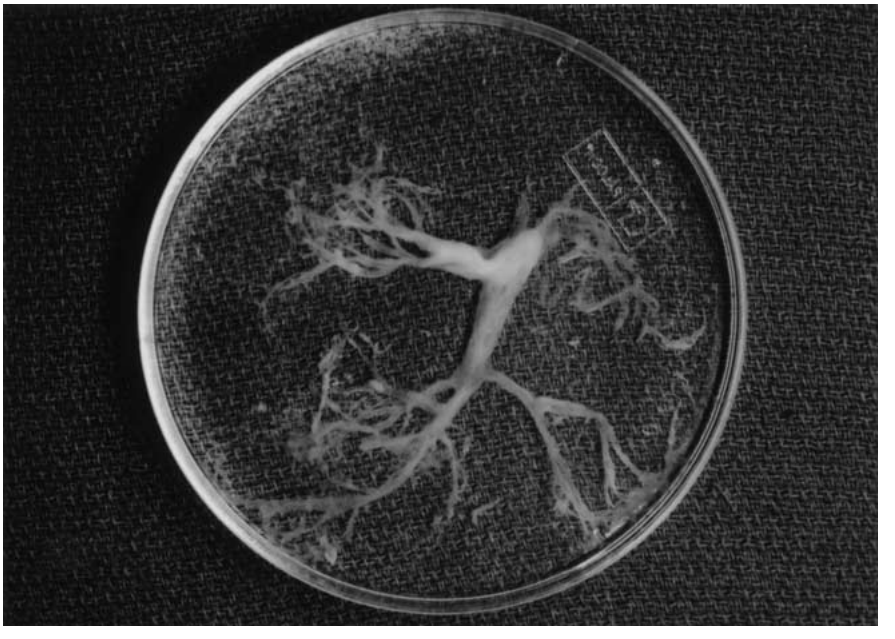

Fig. 1. Bronchial cast expectorated by the patient after a prolonged bout of coughing.

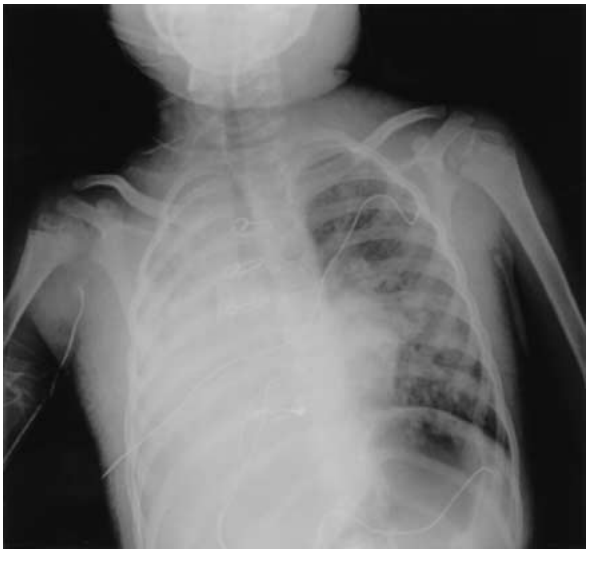

Fig. 2. Baseline chest x-ray, taken after cardiac catheterization and right pulmonary artery stenting, demonstating complete opacification of right hemithorax secondary to atelectasis and air space disease. Interstitial thickening and reticular pattern are noted in the left lung field.

clinically. On recent follow-up, the patient was doing very well, with increased energy and activity and saturations in the low 90s on room air, and has only brought up one cast over the last 9 months since his pacemaker placement. His most recent chest x-ray was also dramatically improved (Fig. 4).

Discussion

Plastic bronchitis has been reported in association with a variety of cardiorespiratory disorders including asthma, cystic fibrosis, allergic bronchopulmonary aspergillosis, bacterial and viral respiratory infections, sickle cell disease with acute chest syndrome, and cyanotic heart disease. The disease is characterized by the development of bronchial casts, which are inflammatory with eosinophilic and fibrinous infiltration (type I, as seen in asthma) or acellular–mucinous (type II, seen in cases of congenital heart disease) [10]. Fontan physiology is increasingly becoming recognized as a risk factor for developing plastic bronchitis [3, 6, 8, 9, 11, 12]. The precise pathophysiology of the disease is unknown, although elevation of pulmonary venous pressure has been ventured as a mechanism in the cases of acellular plastic bronchitis with Fontan circulation. This link between elevated pulmonary venous pressures and cast formation, by an apparent trigger of mucin secretion by the goblet cells of the respiratory mucosa, has been proposed by Seear [11]. Others have suggested endobronchial lymphatic leakage to play a role in cast formation [4, 5], but the exact mechanism is still not understood.

To date no standard treatment is recognized, although several reports have reported some success with a variety of agents, including tissue plasminogen activator, urokinase, steroids, mucolytic agents, and anticoagulation [3, 8]. Previous authors have alluded to the importance of A-V synchrony and optimizing hemodynamics in this disease [9, 11], but to our knowledge this is the first report that clearly shows reestablishment of A-V synchrony as a successful treatment for this disorder.

Fontan physiology predisposes patients to unique complications including pleural effusions, pro-

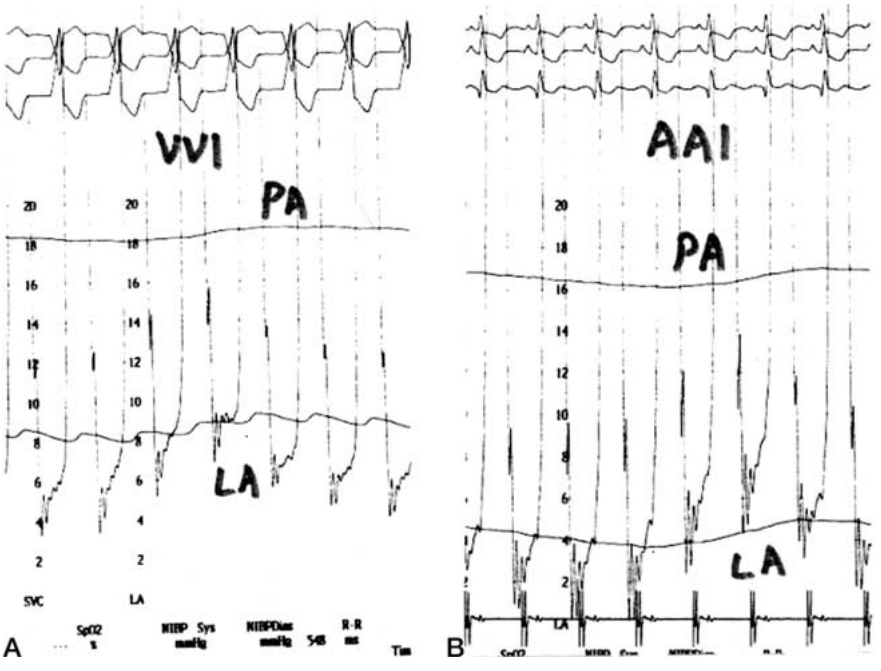

Fig. 3. **A** Baseline hemodynamics with ventricular (VVI) pacing at 110 beat/min, with the patient mechanically ventilated. The top pressure tracing demonstrates an elevated mean pulmonary artery (PA) pressure of 18 mmHg. The lower tracing shows an elevated left atrial (LA) mean pressure of 8 mmHg. Also seen is the left ventricular phasic pressure tracing, revealing an end-diastolic pressure of 6 mmHg. **B** Atrial (AAI) pacing at 110 beats/min and otherwise the same conditions as in A. The mean PA pressure decreased to 16 mmHg. The mean LA pressure decreased to 4 mmHg and the left ventricular end-diastolic pressure decreased to 4 mmHg. *AAI*, atrial mode of pacing; *WI*, ventricular mode of pacing; *LA*, left atrial mean pressure.

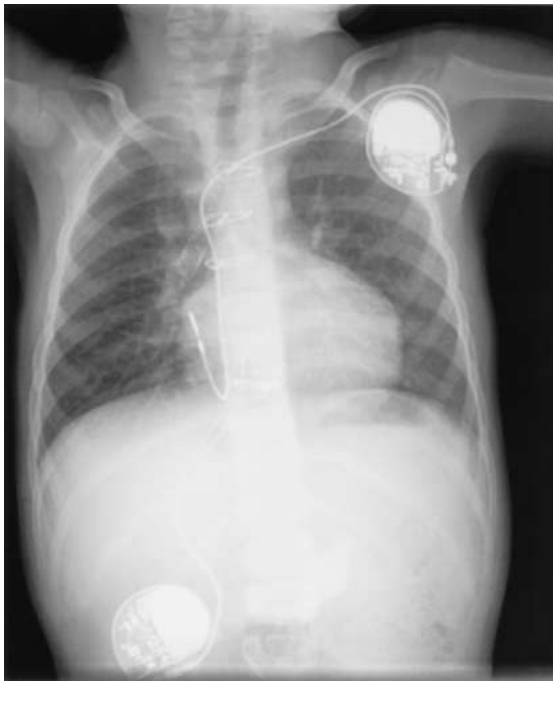

Fig. 4. Follow-up chest x-ray, 2 months after implantation of AAI pacemaker, demonstrating marked improvement, with basically clear lung fields and only mild residual scarring/atelectasis and the right lower base.

tein losing enteropathy, rhythm disturbances, somatic growth retardation, thromboembolic complications, and plastic bronchitis. Atrial pacing has also been proposed as a treatment for Fontan patients with protein-losing enteropathy and sinus node dysfunction [2]. The mechanism of this disorder is also not completely understood but thought to be related to derangement of GI tract hemodynamics caused by the Fontan circulation.

Up to 25% of left ventricular filling occurs with atrial contraction in late diastole [7] in patients with normal anatomy in sinus rhythm. We suspect that in patients with Fontan physiology loss of A-V synchrony will result in even greater decreases in LV filling and cardiac output. Our patient's cardiac output increased by approximately 50% once A-V synchrony was reestablished with atrial pacing, and it is well known that atrial arrhythmias are poorly tolerated in Fontan patients.

The importance of A-V synchrony is not a new concept with regard to the Fontan circulation. Normal sinus rhythm was one of the original criteria sited for patients to be a candidate for the Fontan operation. Our report further demonstrates the importance of optimal hemodynamics in patients with Fontan physiology. More specifically, we believe that the maintenance of A-V synchrony is of paramount importance for patients with Fontan physiology and may prevent long-term complications, such as plastic bronchitis.

Conclusion

Our case demonstrates that acellular plastic bronchitis in a patient with Fontan physiology and A-V

dysynchrony is treatable by restoring A-V synchrony and thereby optimizing hemodynamics. We could infer further that A-V pacing may be beneficial to Fontan patients with sinus node dysfunction, and that ventricular or VVI pacing may be contraindicated in these patients.

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