Cardiology in the Young (2014), **24**, 739–740
doi:10.1017/S1047951113001066
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Brief Report

Severe airway obstruction from a bronchial cast after cardiac transplantation

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Abstract Plastic bronchitis is a rare and difficult to treat disease process in patients with congenital heart disease. Cardiac transplantation has been used increasingly to reverse this process, especially in single ventricle physiology. This case report demonstrates a foreseeable complication after cardiac transplantation in such a patient.

Keywords: Plastic bronchitis; cardiac transplant; single ventricle

Received: 29 May 2013; Accepted: 30 June 2013; First published online: 23 July 2013

PLASTIC BRONCHITIS IS A KNOWN BUT RARE complication in patients after Fontan palliation. It is a sign of failing Fontan physiology, even with preserved ventricular function.[1–3] Plastic bronchitis can lead to severe obstructive airway disease requiring removal of obstructive casts through bronchoscopy.[4] Orthotopic cardiac transplantation has been considered a treatment option for refractory cases.[1,5,6] We describe a unique situation in which a patient with refractory plastic bronchitis and failing Fontan physiology underwent a technically successful cardiac transplant but developed severe obstructive airway disease secondary to a large bronchial cast in the immediate post-operative period. To our knowledge, this is the only reported case of its kind in the literature.

Case report

We present the case of a 12-year-old boy with congenital heart disease in the form of double-inlet left ventricle, right atrioventricular valve atresia, transposed great arteries, severe aortic outflow obstruction, and aortic coarctation. He was status post staged palliation culminating in a fenestrated lateral caval Fontan. He developed plastic bronchitis several years later, which was refractory to all medical therapies and to thoracic duct ligation. Therefore, he underwent cardiac transplantation for failing Fontan physiology.

A day after cardiac transplantation, the patient developed severe respiratory acidosis (arterial pH of 7.04 and $pCO_2 > 100$ mmHg). Severe obstructive airway disease secondary to bronchial casts was highly suspected. Emergent bronchoscopy revealed a large bronchial cast, which was promptly removed, and the respiratory acidosis quickly resolved. Subsequently, inhaled tissue-type plasminogen activator therapy was initiated. He developed no further significant bronchial casts or bleeding complications. A total operating room time of 8 hours for transplantation was likely a risk factor for the development of bronchial casts. The patient is now 3.5 years status post cardiac transplantation and has had no recurrence of plastic bronchitis.

Discussion

Plastic bronchitis is an exceedingly challenging disease to treat.[3,7] It has particularly high mortality and morbidity associated with it in the setting of patients with congenital heart disease, especially among those with Fontan circulation.[8]

Case reports and series have shown that both inhaled tissue-type plasminogen activator and direct

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https://doi.org/10.1017/S1047951113001066 Published online by Cambridge University Press

application of this medication during bronchoscopy may be effective treatments for plastic bronchitis.[8–10] Treatments for plastic bronchitis have been derived from anecdotal evidence owing to its low incidence. Plastic bronchitis has a relapsing nature and high association with morbidity in patients with Fontan circulation. Complete elimination of this condition once it develops is exceedingly difficult, if not impossible in Fontan patients, and treatment is aimed at controlling its severity and frequency. Cardiac transplantation does appear to be effective in eliminating plastic bronchitis in patients with failing Fontans.[1,5,6] Data on the long-term outcomes in these patients is lacking. Despite limited experience with cardiac transplantation for refractory plastic bronchitis, it has shown promise and will likely be an increasingly used treatment for this type of failing Fontan circulation.

This case illustrates that, although resolution of plastic bronchitis after orthotopic cardiac transplantation is likely, severe obstructive airway disease from bronchial casts can still occur in the immediate post-transplant period. Therefore, intensive care physicians, cardiologists, and surgeons alike should be aware of this and proper pre-emptive therapies should be initiated. This should include good respiratory clearance therapy, bronchodilators, and inhaled tissue-type plasminogen activators. Even with these therapies, bronchial casts could occur in the post-operative period. Therefore, a high index of suspicion for severe obstructive airway disease secondary to bronchial cast formation is necessary if respiratory acidosis develops or if the physical exam, chest X-ray, or ventilator mechanics are consistent with severe obstructive airway disease. In this setting, an urgent bronchoscopy with prompt removal of the obstructive casts is paramount.

Conclusions

Plastic bronchitis is a rare but potentially life-threatening disease associated with failing Fontan physiology. Patients with plastic bronchitis undergoing cardiac transplantation are at risk for severe obstructive airway disease secondary to bronchial casts post-operatively. Initiation of empiric inhaled tissue-type plasminogen activator immediately after cardiac transplantation may prevent cast formation and severe obstructive airway disease. In addition, if respiratory acidosis occurs post-operatively, severe obstructive airway disease secondary to bronchial casts should be suspected and bronchoscopy with removal is warranted.

Acknowledgement

None.

Financial Support

This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest

None.

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https://doi.org/10.1017/S1047951113001066 Published online by Cambridge University Press