Cardiopulmonary exercise testing in Fontan patients with and without isomerism (heterotaxy) and patients with ciliary dyskinesia and structurally normal hearts

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Introduction

Isomerism, also known as heterotaxy, is a clinical entity that impacts multiple organ systems both anatomically and functionally. The airways and lungs have been demonstrated to be impacted in a great number of these patients, leading to increased sinopulmonary symptoms, increased need for oxygenation, and increased postoperative ventilatory support. Additionally, these patients often have congenital heart disease requiring Fontan palliation. What has not been previously described, and is the focus of this study, are the results of cardiopulmonary exercise testing in those who have undergone Fontan palliation with and without isomerism. Comparisons were also made to those with primary ciliary dyskinesia as many patients with isomerism have ciliary dyskinesia.

Methods

Patients having undergone cardiopulmonary exercise testing were identified. Patients with isomerism having undergone Fontan palliation were identified from this cohort of patients who had undergone cardiopulmonary exercise testing. The same number of Fontan patients without isomerism were then identified as were the same number of healthy individuals and individuals with primary ciliary dyskinesia but no congenital heart disease.

Results

A total of 68 patients were included in this study, 17 in each of the four groups. Cardiopulmonary exercise testing yielded the best results in healthy patients. All Fontan patients demonstrated mixed pulmonary disease although those with isomerism had greater FVC and FEV1. Exercise times did not differ although peak oxygen consumption was greater in those with isomerism. Those with ciliary dyskinesia had only obstructive pulmonary disease and had the lowest FEF25-75 between all groups. Those with isomerism had a lesser degree of obstructive pulmonary disease when compared to those with primary ciliary dyskinesia.

Conclusion

Fontan patients with and without isomerism have relatively subtle differences in their cardiopulmonary exercise testing, with both groups demonstrating restrictive lung disease. Fontan patients without isomerism tend to be more similar to the primary ciliary dyskinesia group rather than the Fontan patients without isomerism in regards to obstructive lung disease. The results are likely limited by selection bias and highlight the need for multicenter efforts to characterize cardiopulmonary exercise testing in those with isomerism.