



ORIGINAL CLINICAL SCIENCE

Cardiac transplantation in children with Down syndrome, Turner syndrome, and other chromosomal anomalies: A multi-institutional outcomes analysis

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KEYWORDS:

pediatric cardiac transplantation;
chromosomal anomalies;
Down syndrome;
Turner syndrome;
DiGeorge syndrome

BACKGROUND: The purpose of this study was to describe the prevalence, characteristics, and outcomes in pediatric patients with chromosomal anomalies (CA) undergoing orthotopic heart transplantation (OHT).

METHODS: A query of the database of the Pediatric Health Information System, a large administrative and billing database of 43 tertiary children's hospitals, was performed for the Years 2004 to 2016. Pediatric patients who received OHT were analyzed based on presence and type of CA. CA analyzed included: Down syndrome (DS); Turner syndrome (TS)/gonadal dysgenesis; conditions due to anomaly of unspecified chromosome; autosomal deletion; microdeletion; and autosomal anomaly. Healthcare-associated charge analysis during hospitalization for OHT and survival after OHT were assessed.

RESULTS: A total of 3,080 hospitalizations were identified in which OHTs were performed. Of these OHTs, 64 (2.1%) were performed in patients with a concomitant diagnosis of CA. The presence of CA did not confer a higher risk of in-hospital mortality after OHT (odds ratio 1.2 [0.5 to 3.2], $p = 0.651$). Differences in in-hospital mortality between different types of CA, including DS and TS, did not reach statistical significance. Survival at 1-year post-OHT was similar in patients with CA compared to those without CA ($p = 0.248$). Length of stay after OHT was longer in patients with CA: 76 (interquartile range [IQR] 76 to 142 days vs 49 [IQR 21 to 98] days) ($p < 0.001$), respectively. Overall adjusted hospital charges were significantly higher in the CA group: \$1.2 million (IQR \$740,000 to \$2.2 million) vs \$792,000 (IQR \$425,000 to \$1.5 million) ($p < 0.001$), respectively.

CONCLUSIONS: CA is present in ~2% of pediatric patients undergoing OHT. The presence of CA was not associated with increased mortality in pediatric patients undergoing OHT. Limitations of this study include the small number of patients available for analysis and a likely highly selective cohort of patients with CA.

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Cardiac transplantation is the definitive treatment for pediatric patients with end-stage heart failure. Due to the limited availability of donor hearts, patients undergo a

rigorous evaluation before being listed for cardiac transplantation. Listing criteria, as endorsed by the International Society for Heart Lung Transplantation (ISHLT), include evaluation of a patient's cardiopulmonary status, comorbidities, and psychosocial factors that may affect graft/patient survival.¹ Despite a relatively high prevalence within the congenital heart disease cohort,^{2,3} few data are available with regard to orthotopic heart transplantation (OHT) outcomes in patients with chromosomal anomalies (CA). Consideration of CA is not specifically addressed in the 2016 ISHLT listing guidelines.

Currently, little is available to assist cardiac transplant evaluation for patients with CA other than inferential clinical analysis and, perhaps, anecdotal experience. It is unknown whether these patients have a significantly different outcome after cardiac transplantation compared to those without CA. Due to the paucity of this information, we performed a multi-institutional, retrospective query of the Pediatric Health Information System (PHIS) database. Our intent was to study the effect of CA on mortality and associated healthcare utilization after OHT.

Methods

Data source

Data for this study were obtained from the PHIS database, an administrative database that contains inpatient, emergency department, ambulatory surgery, and observation data from 48 not-for-profit, tertiary-care pediatric hospitals in the United States. These hospitals are affiliated with the Children's Hospital Association (Overland Park, KS), a business alliance of children's hospitals. Data quality and reliability are assured through a joint effort between the Children's Hospital Association and participating hospitals. For the purposes of external benchmarking, participating hospitals provide discharge/encounter data that include demographics, diagnoses, procedures, and charges. Data are de-identified at the time of data submission and are subjected to a number of reliability and validity checks before being included in the database. For this study, only data from hospitals that performed OHTs ($n = 30$) were included. Due to the de-identified nature of the PHIS, this study was exempt from review by the institutional review board of Children's Hospital of Philadelphia.

Study population and design

We performed a multi-institutional, retrospective query of the PHIS database for all hospitalizations involving patients ≤ 21 years of age who underwent OHT between January 2004 and March 2016. The primary outcome measure was in-hospital mortality after OHT. Secondary measures included follow-up survival, hospital length of stay (LOS), and hospital charges.

Variables/definitions

All procedural and diagnosis codes of the *International Classification of Diseases, Ninth Revision, Clinical Modification* (ICD-9) were reviewed. OHT was defined by the presence of ICD-9 Code 37.51 and was the basis for inclusion in the study. CA were defined as the presence of ICD-9 codes representing: chromosomal anomaly (Down syndrome: 758.0; Turner syndrome/gonadal

dysgenesis: 758.6; other conditions due to sex chromosome anomalies: 758.81; conditions due to anomaly of unspecified chromosome: 758.9); autosomal deletion (autosomal deletion syndrome: 758.3; other autosomal deletions: 758.39); microdeletion (DiGeorge syndrome: 279.11; other microdeletions: 758.33); and autosomal anomaly (other conditions due to autosomal anomalies: 758.5).

Other variables were taken directly from the PHIS data elements including: patient demographics data; mortality; LOS; OHT within 24 hours of admission; and adjusted hospital charge data. Hospital charges are adjusted within the PHIS by the Centers for Medicare and Medicaid Services wage-to-price index for the hospital's location and are categorized as: clinical; pharmacy; imaging; laboratory; supply; and other (primarily room and nursing) charges. The total adjusted charge is equal to the arithmetic sum of all 5 charge categories.

A unique patient identifier is assigned within the PHIS for each patient at a given hospital, allowing for longitudinal tracking of individual patients for visits occurring at the same hospital. Mortality was defined by the presence of a discharge mortality flag within the PHIS and was recorded at the index hospitalization as well as at any subsequent clinical encounter that occurred during follow-up for survival analysis. Thus, patients in this study were longitudinally followed until their last known clinical encounter or until death. Data were censored if a patient did not have any follow-up at the index hospital after the index hospitalization.

Statistical analysis

Given the data were not normally distributed, descriptive statistics were reported using median and interquartile range (IQR) for continuous variables and proportions for categorical variables. Univariable analysis of categorical variables and mortality was performed using Pearson's chi-square or Fisher's exact test, as appropriate. Continuous variables were analyzed using the Mann-Whitney U -test. Multivariable analyses were performed utilizing binary logistic regression analysis. Models were controlled for presence of CA, presence of cardiomyopathy, OHT within 24 hours of hospital admission, gender, age at transplantation, and age of patient (infant, child, or adolescent). Follow-up survival analysis was performed using Kaplan-Meier survival estimates. Two-tailed test of significance was used in all statistical analyses, with statistical significance defined as $p < 0.05$, and all analyses were performed using SPSS Statistics version 19 (IBM SPSS Corp., Armonk, NY).

Results

Between January 2004 and March 2016, 3,080 hospitalizations across 30 hospitals were identified in which an OHT was performed. A total of 64 (2.1%) OHTs were performed in patients with a concomitant diagnosis of CA. [Table 1](#) demonstrates the demographic comparison between the CA and no-CA transplant recipients. Both gender and OHT within 24 hours of admission were statistically different between the patients with and without CA. Patients' demographics by CA type are listed in [Table 2](#). Although there were no statistical differences between these groups, small sample sizes limit these data. The frequencies of specific CA are displayed in [Figure 1](#). By category, incidence of CA included chromosomal anomaly in 31 (1.0%), autosomal deletion in 8 (0.3%), microdeletion in 14 (0.5%), and autosomal anomaly in 11 (0.4%).

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