Perioperative Management of Patients With DiGeorge Syndrome Undergoing Cardiac Surgery

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Objective: **DiGeorge syndrome is a genetic disorder with multisystem involvement resulting in craniofacial and cardiac anomalies and parathyroid and immune system dysfunction. This study describes perioperative management of a large cohort of patients with DiGeorge syndrome undergoing cardiac surgery.**

Design: **Retrospective cohort study.**

Setting: **Major academic tertiary institution.**

Participants: **The medical records of patients diagnosed with DiGeorge syndrome and undergoing cardiac surgery at this institution, from January 1, 1976, to July 31, 2012, were reviewed for phenotypic characteristics and intraoperative and postoperative complications, with specific attention to hemodynamic instability, perioperative perturbations of plasma calcium homeostasis, and airway difficulty.**

Interventions: **None.**

Measurements and Main Results: **Sixty-two patients underwent 136 cardiac surgical procedures; 47 patients (76%) had multiple operations. Sternotomies for reoperations often were complex (8 complicated by vascular injury or difficulty achieving hemostasis and 5 requiring bypass before sternotomy). Two patients had persistent hypocalcemia intraoperatively, requiring infusion of calcium chloride, and hypocalcemia developed postoperatively in 8 patients. Prolonged mechanical ventilation (>24 hours) was required after 48 procedures (35%), and 25 (18%) required prolonged inotropic support (>72 hours). Infectious complications occurred after 31 procedures (23%). There was no in-hospital or 30-day mortality.**

Conclusions: **Patients with DiGeorge syndrome often have complex cardiac anomalies that require surgical repair. The postoperative course is notable for the frequent need for prolonged respiratory and hemodynamic support. Patients can develop hypocalcemia and may require calcium supplementation. Immunodeficiencies may be associated with the increased rate of postoperative infections and may dictate the need for specific transfusion management practices.**

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KEY WORDS: anesthesia, cardiovascular surgical procedures, DiGeorge syndrome, hypocalcemia, immunosuppression, infection

DIGEORGE SYNDROME (DGS), or 22q11.2 deletion syndrome, is a multisystem disorder associated with congenital heart disease, craniofacial abnormalities, developmental delays, and disorders of the endocrine, immune, and hematologic systems.[1,2] It is inherited in an autosomal dominant fashion, but most deletions (>90%) occur as a de novo mutation.[3] Patients typically have cyanotic heart disease secondary to conotruncal heart defects (malformations of the cardiac outflow tracts) such as tetralogy of Fallot, pulmonary atresia with ventricular septal defect, truncus arteriosus, interrupted aortic arch, and ventricular septal defect.[4]

Only a few reports have been published describing the perioperative management of patients with DGS undergoing surgical repair of congenital cardiac anomalies.[4–10] Because this syndrome is uncommon, case series and case reports are required to provide information regarding perioperative challenges in the management of patients with DGS. This institution has long experience in caring for patients with congenital heart disease and a comparatively large number of DGS patients who have undergone cardiac surgery. The aim of this study was to report the perioperative course of patients with DGS who had had cardiac surgery for congenital abnormalities at this large tertiary center.

METHODS

This retrospective medical record review received approval from the Institutional Review Board. The authors included only patients who provided authorization for research use of their medical records (historically >95% of the patients).[11] This research was conducted in compliance with the World Medical Association Declaration of Helsinki.

A computerized search of medical record databases at this institution, from January 1, 1976, to July 31, 2012, was conducted to identify all patients with a genetic or clinical diagnosis of DGS. The records of identified patients were reviewed further to identify those who underwent cardiac surgery at this institution (charts reviewed by TY, FS, and RJH). Patients who underwent cardiac surgery at another institution were not included. Patients who underwent minor cardiac surgical procedures such as cardiac catheterization (both diagnostic and interventional) and insertion of pacemakers under sedation were not included. However, those who underwent placement of pacemakers or implantable cardioverter-defibrillators with epicardial leads that required a sternotomy or thoracotomy under general anesthesia were included.

The genetic diagnosis of DGS was made with the fluorescent in situ hybridization test to identify deletions on chromosome 22q11.2.[12,13] All clinical diagnoses were made by a staff physician specialist in pediatrics, pediatric cardiology, or genetic medicine on the basis of phenotypic characteristics, family history, and presence of multiple congenital heart malformations.

Medical records were reviewed for phenotypic characteristics of DGS: Congenital cardiovascular, craniofacial, and airway anomalies; disorders of the immune, endocrine, gastrointestinal, genitourinary, neurologic, ophthalmologic, skeletal, and hematologic systems; developmental and neuropsychiatric abnormalities; and other comorbid conditions unrelated to DGS. Surgical and anesthetic records were reviewed for intraoperative events such as difficult airway management,

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blood transfusions, interventions to provide hemodynamic support such as inotropic infusions or mechanical interventions such as intra-aortic balloon pump or extracorporeal membrane oxygenation, hypocalcemia, arrhythmias requiring interventions, and specific documentation of difficulty achieving hemostasis or inadvertent vessel injury. Hospital records were reviewed for postoperative complications. Reflecting the typical surgical practice with pediatric cardiac patients, prolonged ventilation was defined as requiring mechanical ventilation for more than 24 postoperative hours and prolonged inotropic support was defined when being extended beyond 72 postoperative hours. Infections were defined as fever (temperature $>38^{\circ}C$); treatment with antibiotics; and positive blood, respiratory, or urine cultures. However, isolated sternal wound infections were considered as infectious complications even if not accompanied by fever.

Descriptive summaries of demographic, epidemiologic, and perioperative complications and other data were performed and are reported as medians or frequencies (percentages). Because of the prolonged timespan of this study, additional analysis was performed comparing outcomes of surgeries performed during the early (before the year 2000) and later epoch (year 2000 and after) in regard to duration of postoperative mechanical ventilation, inotropic support, and intensive care unit days. The frequency of infections between the two epochs also was compared.

To review the current knowledge regarding anesthetic complications in patients with DGS undergoing cardiac surgery, the authors performed a literature search of MEDLINE (January 1, 1946 to July 31, 2012) and EMBASE (January 1, 1988 to July 31, 2012) databases using the following text words: *22q11 deletion syndrome* or *DiGeorge syndrome*, *anesthesia*, *anesthetics*, *intraoperative*, *preoperative*, and *postoperative*. Published articles and selected bibliographies were reviewed for relevancy.

RESULTS

One hundred thirty-one patients with DGS were evaluated at this institution from January 1, 1976, to July 31, 2012. Of these, 62 patients underwent 136 cardiac surgical procedures. Of these, 64 cases (47%) were performed in the contemporary epoch (2000 or later) and 72 (53%) cases in the early epoch (47 cases during the 1990s, 25 cases before 1990). From 2000 to 2012 there were 33,119 cardiac surgeries; thus, DGS cardiac surgical patients accounted for 0.2% of the contemporary practice. Characteristics and comorbid conditions of patients in this cohort are summarized in Table 1. Diagnosis of DGS was made before the first cardiac surgery in 18 (29%) patients and after the first cardiac surgery in 44 (71%) patients. Fifty-six patients (90%) had cyanotic conotruncal heart abnormalities, and 47 patients (76%) had multiple cardiac surgical procedures. Other congenital anomalies were common.

Surgical characteristics and intraoperative complications are summarized in Table 2. The three most common surgical procedures were systemic-to-pulmonary shunts, valvular operations, and unifocalization procedures (reincorporation of the aortopulmonary collaterals into the pulmonary vasculature for treatment of pulmonary atresia). Ninety-one procedures were performed using cardiopulmonary bypass. All patients were weaned from cardiopulmonary bypass. However, one patient who underwent repair of pulmonary atresia required a return to bypass for reopening of a ventricular septal defect because of persistently high right ventricular pressure. Six patients underwent eight procedures with difficulty achieving hemostasis and/or inadvertent vessel injury during sternotomy. None of these

Table 1. Characteristics and Comorbid Conditions in Patients with DiGeorge Syndrome

Patient Characteristics and Comorbid Conditions	Total Patients (N = 62)
Male sex	32 (52)
ASA status ≥3	62 (100)
Diagnosis	
Genetic	60 (96.8)
Phenotypic	2 (3.2)
Age at diagnosis, y[*]	2 (0.1-14)
Age at first cardiac surgery at this institution, y	1 (0.2-7)
Cardiovascular disease	62 (100)
Cyanotic heart disease	
Tetralogy of Fallot	16
Pulmonary atresia and VSD	21
Pulmonary stenosis and VSD	5
Truncus arteriosus	4
Interrupted aortic arch or coarctation of the aorta	8
Atrioventricular canal defect	1
Double-outlet right ventricle, pulmonary stenosis, VSD	1
Acyanotic heart disease	
VSD, ASD, PDA	6
Valvular heart disease	5
Vascular ring or major arterial malformations (aortic arch, anomalous subclavian, carotid)	17
Airway and ear, nose, and throat disease	32 (52)
Velopharyngeal incompetence or insufficiency	9
Upper airway anomalies[†]	9
Laryngomalacia, tracheomalacia, or bronchomalacia	3
Recurrent acute otitis media or chronic otitis media	19
Micrognathia or retrognathia	1
Hearing loss (sensorineural, conductive, or mixed)	10
Immune-related disease	21 (34)
Recurrent infections (UTI, pneumonia, URTI)	14
Low T-cell count or impaired function	7
Autoimmune diseases[‡]	5
Immunoglobulin deficiency	3
Absent thymus or thymic hypoplasia	3[§]
Endocrine disease	29 (47)
Hypocalcemia with or without hypoparathyroidism	24
Thyroid disorders	9
Gastrointestinal disease	15 (24)
Gastroesophageal reflux disease	11
Dysmotility or dysphagia	6
Hernia (umbilical, inguinal, or diaphragmatic)	3
Genitourinary disease	3 (5)
Unilateral renal agenesis or unilateral absent kidney/congenital hydronephrosis	3
Vesicoureteral reflux	1
Ophthalmic disease	9 (15)
Strabismus	4
Refractory errors or astigmatism	5
Musculoskeletal disease	13 (21)
Scoliosis or other spine anomalies	13
Microcephaly and plagiocephaly	1
Hematologic disease	8 (13)
Thrombocytopenia	8
Neurologic disease	6 (10)
Recurrent seizures (>1 episode) (with or without hypocalcemia)	6

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