Published in final edited form as:

World J Pediatr Congenit Heart Surg. 2011 April 1; 2(2): 278–286. doi:10.1177/2150135110397670.

Heterotaxy: Lessons Learned about Patterns of Practice and Outcomes from the Congenital Heart Surgery Database of the Society of Thoracic Surgeons

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Abstract

Background—“Heterotaxy is synonymous with ‘visceral heterotaxy’ and ‘heterotaxy syndrome’. Heterotaxy is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as ‘situs solitus’, nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as ‘situs inversus’.” The purpose of this manuscript is to review the data about heterotaxy in the STS Congenital Heart Surgery Database.

Methods—We examined all index operations in the STS Congenital Heart Surgery Database over 12 years from January 1, 1998 - December 31, 2009, inclusive. This analysis resulted in a cohort of 77,153 total index operations. Of these 77,153 index operations, 1,505 (1.95%) were

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performed in patients with heterotaxy. Of the 1,505 index operations performed in patients with heterotaxy, 1,144 were in patients with “asplenia” and 361 were in patients with “polysplenia”.

Results—In every STS–EACTS Congenital Heart Surgery Mortality Category (2009) (STS–EACTS Category), discharge mortality is higher in patients with heterotaxy compared to patients without heterotaxy. Discharge mortality after Systemic to pulmonary artery shunt is 6.6% in a cohort of all Single Ventricle patients except those with heterotaxy, while it is 10.8% in Single Ventricle patients with heterotaxy. Discharge mortality after Fontan is 1.8% in a cohort of all Single Ventricle patients except those with heterotaxy, while it is 4.2% in Single Ventricle patients with heterotaxy.

Conclusions—The STS Database is largest Congenital Heart Surgery Database in North America. This review of data from the STS Congenital Heart Surgery Database allows for unique documentation of practice patterns and outcomes. From this analysis, it is clear that heterotaxy is a challenging problem with increased discharge mortality in most subgroups.

Keywords

heterotaxy syndrome; isomerism; asplenia; polysplenia; database; congenital heart disease; outcomes; cardiac surgery; results of treatment

Background

The Congenital Heart Surgery Database of the Society of Thoracic Surgeons (STS) is the largest database in North America that tracks the outcomes of pediatric and congenital heart surgery [1]. The Congenital Heart Surgery Database of The European Association for Cardio-Thoracic Surgery (EACTS) is the largest database in Europe that tracks the outcomes of pediatric and congenital heart surgery [2, 3]. The International Nomenclature Committee for Pediatric and Congenital Heart Disease (ISNPCHD) offers the following definition for heterotaxy [4]:

“Heterotaxy is synonymous with ‘visceral heterotaxy’ and ‘heterotaxy syndrome’. Heterotaxy is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as ‘situs solitus’, nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as ‘situs inversus’.”

It is helpful to understand that (1) asymmetry of the thoracic and abdominal viscera is the usual or normal situation, and (2) heterotaxy syndrome includes patients with an unusual degree of thoracic and abdominal visceral symmetry. (The term “viscera” is defined as the internal organs of the body, specifically those within the chest or abdomen.) Although many patients with heterotaxy undergo univentricular palliation, heterotaxy is not a surrogate for “single ventricle”. Some patients with heterotaxy will undergo univentricular palliation while others will undergo biventricular repair.

We have recently published data from the STS Congenital Heart Surgery Database about “Lessons Learned about Patterns of Practice and Outcomes” related to Atrioventricular Septal Defects [5] and Transposition of the Great Arteries [6]. The purpose of this manuscript is to review the data about heterotaxy in the STS Congenital Heart Surgery Database. This manuscript will first review the system of nomenclature and classification used in the STS Congenital Heart Surgery Database for heterotaxy and will then present the data contained in the STS Congenital Heart Surgery Database about heterotaxy.

Material and Methods

Nomenclature

The STS Congenital Heart Surgery Database, The EACTS Congenital Heart Surgery Database, and The Japan Congenital Cardiovascular Surgery Database (JCCVSD) all use the same nomenclature and database definitions and standards. The nomenclature used by these three databases is the International Paediatric and Congenital Cardiac Code (IPCCC) that was developed and is maintained by the ISNPCHD (www.ipccc.net) [7]. These three databases use the version of the IPCCC derived from the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and the STS (www.ipccc.net) [2, 7, 8]. On January 1, 2010, these three databases upgraded to a new version of specifications for the definitions and standards in the database (Version 3.0) [9]. This new version of specifications uses ten terms related to heterotaxy: seven diagnostic terms and three syndromes. In the next section of this manuscript, we will present the terms and definitions related to heterotaxy utilized in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database, and the JCCVSD.

Terminology and Definitions

As stated above, the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database, and the JCCVSD all use ten terms related to heterotaxy: three syndromes and seven diagnostic terms [9].

Syndromes—The following terms are contained within the Syndromes Short List of the STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and the JCCVSD:

1. Heterotaxy syndrome
2. Heterotaxy syndrome, Asplenia syndrome
3. Heterotaxy syndrome, Polysplenia syndrome

Diagnostic terms—The following diagnostic terms are contained within the Diagnosis Short List of the STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and the JCCVSD:

1. Single ventricle, Heterotaxia syndrome
2. Atrial Isomerism, Left {CAN NOT BE PRIMARY DIAGNOSIS}
3. Atrial Isomerism, Right {CAN NOT BE PRIMARY DIAGNOSIS}
4. Dextrocardia {CAN NOT BE PRIMARY DIAGNOSIS}
5. Levocardia {CAN NOT BE PRIMARY DIAGNOSIS}
6. Mesocardia {CAN NOT BE PRIMARY DIAGNOSIS}
7. Situs inversus {CAN NOT BE PRIMARY DIAGNOSIS}

It should be noted that the term “**Single ventricle, Heterotaxia syndrome**” has been in used by the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database for over a decade [8, 10]. The term “Heterotaxia syndrome” is synonymous with the following three “heterotaxy”, “visceral heterotaxy” and “heterotaxy syndrome”. In future versions of the STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and the JCCVSD, it is very likely that the term Single ventricle,

Heterotaxia syndrome” will be replaced by the term Single ventricle, Heterotaxia syndrome” will be replaced by the term “Single ventricle, Heterotaxy”.

Definitions—The following definitions of the above terms are from the ISNPCHD [4, 11]:

Heterotaxy syndrome

“Heterotaxy is synonymous with ‘visceral heterotaxy’ and ‘heterotaxy syndrome’. Heterotaxy is defined as an abnormality where the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy does not include patients with either the expected usual or normal arrangement of the internal organs along the left-right axis, also known as ‘situs solitus’, nor patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as ‘situs inversus’.” [4]

Heterotaxy syndrome, Asplenia syndrome

“‘Asplenia syndrome’ can be defined as a subset of heterotaxy with components of bilateral right-sidedness, usually associated with absence of the spleen.” [4]

Heterotaxy syndrome, Polysplenia syndrome

“‘Polysplenia syndrome’ can be defined as a subset of heterotaxy with components of bilateral left-sidedness, usually associated with multiple spleens.” [4]

Single ventricle, Heterotaxia syndrome

The functionally univentricular heart (also known as ‘Single ventricle’) is defined as a spectrum of cardiac malformations in which the entire ventricular mass is functionally univentricular; in other words, whenever only one ventricle is capable, for whatever reason, of supporting either the systemic or the pulmonary circulation. ‘Single ventricle, Heterotaxia syndrome’ is a functionally univentricular heart in the setting of heterotaxy. [4, 11]

Atrial Isomerism, Left {CAN NOT BE PRIMARY DIAGNOSIS}

“Isomerism of the left atrial appendages is a subset of heterotaxy where the atrial appendages on both sides of the body have the appearance of the morphologically left atrial appendage.” [4]

Atrial Isomerism, Right {CAN NOT BE PRIMARY DIAGNOSIS}

“Isomerism of the right atrial appendages is a subset of heterotaxy where the atrial appendages on both sides of the body have the appearance of the morphologically right atrial appendage.” [4]

Dextrocardia {CAN NOT BE PRIMARY DIAGNOSIS}

Dextrocardia is defined as a right-sided ventricular mass. In other words, the heart is predominantly in the right hemithorax.

Levocardia {CAN NOT BE PRIMARY DIAGNOSIS}

Levocardia is defined as a left-sided ventricular mass. In other words, the heart is predominantly in the left hemithorax.

Mesocardia {CAN NOT BE PRIMARY DIAGNOSIS}

Mesocardia is defined as a central/midline ventricular mass. In other words, the heart is midline.

Situs inversus {CAN NOT BE PRIMARY DIAGNOSIS}

Situs inversus is synonymous with “Total mirror imagery” and is defined as complete mirror-imaged arrangement of the internal organs along the left-right axis.

STS Congenital Heart Surgery Database

The STS Congenital Heart Surgery Database is the largest database in North America dealing with congenital cardiac malformations. In a previous publication in *The World Journal for Pediatric and Congenital Heart Surgery* (WJPCHS), we described the scope of the STS Congenital Heart Surgery Database [5]. We will now briefly update this information.

The STS Congenital Heart Surgery Database has grown annually since its inception, both in terms of the number of participating centers submitting data, and the number of operations analyzed (Figures 1, 2, and 3) [1]. As of October 1, 2010, the STS Congenital Heart Surgery Database currently has 98 Participating Centers: 96 from the United States of America, one from Canada, and one from Japan. The Report of the 2005 STS Congenital Heart Surgery Practice and Manpower Survey, undertaken by the STS Workforce on Congenital Heart Surgery, documented that 122 centers in the United States of America perform pediatric and congenital heart surgery and 8 centers in Canada perform pediatric and congenital heart surgery [12]. Between January 1, 1998 and December 31, 2009, inclusive, the number of cumulative total operations in the STS Congenital Heart Surgery Database is 164,240 [1].

The data presented in this manuscript will be derived from all index operations in the STS Congenital Heart Surgery Database with a date of surgery between January 1, 1998 and December 31, 2009, inclusive. An Index Operation is defined as the first cardiac operation of a hospitalization [1]. A cardiac operation is defined as an operation with the operation type coded as operation type “CPB” or “No CPB Cardiovascular” (CPB = cardiopulmonary bypass) [1].

Mortality data reported in this manuscript is “discharge mortality”, or mortality prior to discharge from the hospital. Due to the descriptive nature of this analysis, formal statistical comparisons were not made between the various cohorts in this analysis. Consequently, p-values have not been provided.

Results**Data from The STS Congenital Heart Surgery Database**

All of the above 10 terms related to heterotaxy listed in the Material and Methods section are in the current version 3.0 [9] of the STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and the JCCVSD. This version 3.0 went live on January 1, 2010 [9]. The time period of the analysis reported in the Results section of this manuscript is all patients in the STS Congenital Heart Surgery Database who had surgery between January 1, 1998 and December 31, 2009, inclusive. The nomenclature used during this time interval only contained the 5 terms related to heterotaxy: two Non-Cardiac Abnormalities and three Diagnoses:

Non-Cardiac Abnormalities

1. Asplenia

2. Polysplenia

Diagnosis

1. Single ventricle, Heterotaxia syndrome
2. Atrial Isomerism, Left
3. Atrial Isomerism, Right

“Asplenia” and “Polysplenia” in this analysis are defined the same as the following terms in the Material and Methods section: “Heterotaxy syndrome, Asplenia syndrome” and “Heterotaxy syndrome, Polysplenia syndrome”. The three diagnoses used during the time interval of this analysis are also defined in the Material and Methods section.

We initially examined all index operations in the STS Congenital Heart Surgery Database over 12 years from January 1, 1998 – December 31, 2009, inclusive, and excluded the following three groups of operations:

1. Operations performed by STS Database Participants with > 15% missing data for the following fields in the STS Congenital Heart Surgery Database: discharge mortality or non-cardiac abnormalities (An STS Database Participant is either a “practice group of cardiothoracic surgeons” or an individual cardiothoracic surgeon.)
2. Individual records of operations missing data for the following fields: discharge mortality or non-cardiac abnormalities
3. Operations performed on patients weighing less than or equal to 2,500 g undergoing PDA ligation as their primary procedure.

This analysis resulted in a cohort of 77,153 total index operations. Of these 77,153 index operations, 1,505 (1.95%) were performed in patients with heterotaxy. Of the 1,505 index operations performed in patients with heterotaxy, 1,144 were in patients with “asplenia” and 361 were in patients with “polysplenia”.

Table 1 documents all index operations in the STS Congenital Heart Surgery Database performed between January 1, 1998 and December 31, 2009, inclusive, and documents the number of operations and the mortality prior to discharge from the hospital after cardiac surgery. This information is reported for all operations and is also reported stratified by the STS-EACTS Congenital Heart Surgery Mortality Category (2009) (or, briefly, the STS-EACTS Category) [13]. This information is reported for the following four groups:

1. All Index Operations in Database
2. All index operation in patients with “Asplenia”
3. All index operation in patients with “Polysplenia”
4. All index operation in patients without “Asplenia” or “Polysplenia”

The data in Table 1 includes all index operations in the STS Congenital Heart Surgery Database over 12 years from January 1, 1998 – December 31, 2009, inclusive, and excludes the following three groups of operations:

1. Operations performed by STS Database Participants with > 15% missing data for the following fields in the STS Congenital Heart Surgery Database: discharge mortality or non-cardiac abnormalities
2. Individual records of operations missing data for the following fields: discharge mortality or non-cardiac abnormalities

3. Operations performed on patients weighing less than or equal to 2,500 g undergoing PDA ligation as their primary procedure.

Table 2 documents all index operations in the STS Congenital Heart Surgery Database performed between January 1, 1998 and December 31, 2009, inclusive, in patients with the diagnosis of any form of “Single Ventricle” with any the following diagnoses:

1. Single ventricle, DILV
2. Single ventricle, DIRV
3. Single ventricle, Mitral atresia
4. Single ventricle, Tricuspid atresia
5. Single ventricle, Unbalanced AV canal
6. Single ventricle, Heterotaxia syndrome
7. Single ventricle, Other

and an operation that falls into one of the following three groups of procedures:

1. All Index Shunt Operations
2. All Index Glenn Operations
3. All Index Fontan Operations

It should be noted that the data in Table 2 does not include patients with the diagnosis of “Hypoplastic left heart syndrome (HLHS)”.

“All Index Shunt Operations” includes the following primary procedures:

1. Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)
2. Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery)
3. Shunt, Systemic to pulmonary, Other

“All Index Glenn Operations” includes the following primary procedures:

1. Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)
2. Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn)
3. Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn)
4. HemiFontan

“All Index Fontan Operations” includes the following primary procedures:

1. Fontan, Atrio-pulmonary connection
2. Fontan, Atrio-ventricular connection
3. Fontan, TCPC, Lateral tunnel, Fenestrated
4. Fontan, TCPC, Lateral tunnel, Nonfenestrated
5. Fontan, TCPC, External conduit, Fenestrated
6. Fontan, TCPC, External conduit, Nonfenestrated
7. Fontan, Other

Table 2 reports documents the number of operations and the mortality prior to discharge from the hospital after cardiac surgery for the above cohorts of patients for the following three groups:

1. All Single Ventricle
2. All Single Ventricle except Heterotaxy
3. Single ventricle, Heterotaxia syndrome

“All Single Ventricle” includes the following primary diagnoses:

1. Single ventricle, DILV
2. Single ventricle, DIRV
3. Single ventricle, Mitral atresia
4. Single ventricle, Tricuspid atresia
5. Single ventricle, Unbalanced AV canal
6. Single ventricle, Heterotaxia syndrome
7. Single ventricle, Other

“All Single Ventricle except Heterotaxy” includes the following primary diagnoses:

1. Single ventricle, DILV
2. Single ventricle, DIRV
3. Single ventricle, Mitral atresia
4. Single ventricle, Tricuspid atresia
5. Single ventricle, Unbalanced AV canal
6. Single ventricle, Other

“Single ventricle, Heterotaxia syndrome” includes the following primary diagnosis:

1. Single ventricle, Heterotaxia syndrome

The data in Table 2 excludes the following two groups of operations:

1. Operations performed by STS Database Participants with > 15% missing data for the following field in the STS Congenital Heart Surgery Database: discharge mortality
2. Individual records of operations missing data for the following field: discharge mortality.

Discussion

The STS Database is the largest Congenital Heart Surgery Database in North America. This review of data from 1,505 operations performed in patients with heterotaxy allows for unique documentation of patterns of practice and outcomes. From this review, we have learned that heterotaxy is a challenging problem with increased discharge mortality in most subgroups.

In every STS–EACTS Congenital Heart Surgery Mortality Category (2009) (STS–EACTS Category), discharge mortality is higher in patients with heterotaxy compared to those without heterotaxy. Discharge mortality after Systemic to pulmonary artery shunt is 6.6% in

a cohort of all Single Ventricle patients except those with heterotaxy, while it is 10.8% in those with heterotaxy. Discharge mortality after Fontan is 1.8% in a cohort of all Single Ventricle patients except those with heterotaxy, while it is 4.2% in those with heterotaxy.

In multiple individual institutional reports and review manuscripts, heterotaxy has been associated with increased operative mortality and morbidity in a variety of cardiac surgical settings. Survival after repair of total anomalous pulmonary venous connection in patients with heterotaxy has consistently been reported in the literature to be far inferior to the results obtained in patients without heterotaxy [14]. However, a recent series from Texas Children's Hospital documented that "At midterm follow-up, heterotaxy patients undergoing total anomalous pulmonary venous return repair have a higher pulmonary vein reoperative rate but a comparable overall survival to nonheterotaxy patients." [14]. The numerous challenges associated with univentricular palliation in patients with heterotaxy syndrome have been summarized as follows: "Traditionally, those patients with heterotaxy who survive initial surgical management have been considered a very high-risk population with respect to an eventual Fontan procedure. In heterotaxy patients, it is particularly important to understand the unique and variable anatomy of the sinus node and conduction system, the potential for occult pulmonary venous obstruction, the tendency for development of atrioventricular valve regurgitation in volume loaded ventricles, and the potential for recurrent or persistent cyanosis because of intrahepatic shunting or pulmonary arteriovenous malformations." [15]. Finally, heterotaxy has also been shown to be a risk factor for mortality after cardiac transplantation [16].

If one considers the amount of data contained in the STS Congenital Heart Surgery Database about patients with heterotaxy, this analysis represents merely the "tip of the iceberg" of potential analyses. Several additional detailed studies could be performed related to patients with heterotaxy using the STS Congenital Heart Surgery Database:

1. The endpoint of this analysis is discharge mortality. More detailed analyses can be performed of the cohorts reported in this manuscript to examine morbidity, including length of stay in the hospital and complications.
2. An additional analysis can be performed focusing on patients with heterotaxy and total anomalous pulmonary venous connection (TAPVC).
3. An additional analysis can be performed focusing on pulmonary venous stenosis in patients with heterotaxy and total anomalous pulmonary venous connection (TAPVC).
4. Additional analyses can be performed comparing outcomes after the various types of systemic to pulmonary artery shunts in patients with heterotaxy.
5. Additional analyses can be performed involving heterotaxy patients who underwent palliative banding of the pulmonary artery as a prelude to univentricular palliation or even biventricular repair.
6. Additional analyses can be performed comparing outcomes after the various types of superior cavopulmonary anastomoses in patients with heterotaxy.
7. Additional analyses can be performed comparing outcomes after the various types of Fontan operations in patients with heterotaxy.
8. Additional analyses to evaluate the role of univentricular palliation versus biventricular repair in patients with heterotaxy [17, 18].
9. Additional analyses can be performed to study the outcomes of patients with heterotaxy after cardiac transplantation.

The purpose of this manuscript is not to make detailed statistical comparisons between various groups. The purpose of this manuscript is also not to create formal statistical models to assess risk or predict outcome. Future analyses may be performed to accomplish these goals. The purpose of this manuscript is simply to obtain a very general and broad view of the scope of information in the STS Congenital Heart Surgery Database about heterotaxy.

The main limitation of this analysis is the main limitation of The STS Congenital Heart Surgery Database: it only allows for analysis of outcomes up to discharge from the hospital. By transforming The STS Congenital Heart Surgery Database into a platform for longitudinal follow-up [19, 20, 21, 22, 23, 24], important information about the intermediate-term and long-term outcomes of patients with heterotaxy undergoing cardiac surgery will become available. Therefore, on January 1, 2010, The STS Congenital Heart Surgery Database began collecting Unique Patient Identifiers compliant with the Health Insurance Portability and Accountability Act (HIPAA) of the Federal government of the United States of America. These Unique Patient Identifiers will allow The STS Congenital Heart Surgery Database to function as a tool for longitudinal follow-up and will lead to the collection of important data about the intermediate-term and long-term status of our patients.

Conclusions

1. The STS Database is largest Congenital Heart Surgery Database in North America.
2. This review of data from the STS Congenital Heart Surgery Database allows for unique documentation of practice patterns and outcomes.
3. From this analysis, it is clear that heterotaxy is a challenging problem with increased discharge mortality in most subgroups.

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Figure 1.

The graph documents the annual growth in the STS Congenital Heart Surgery Database by number of Participating Centers. As of October 1, 2010, the STS Congenital Heart Surgery Database has 98 Participating Centers: 96 from the United States of America, one from Canada, and one from Japan.

Growth in the STS Congenital Heart Surgery Database

Figure 2.

The graph documents the annual growth in the STS Congenital Heart Surgery Database by the number of operations per averaged 4-year data collection cycle. The aggregate report from the Spring 2010 Harvest of the STS Congenital Heart Surgery Database included 98,628 operations performed in the 4-year period of January 1, 2006 through December 31, 2010, inclusive, submitted from 85 centers from North America, 84 from the United States of America and 1 from Canada. One Japanese center also submits data; however, these Japanese data are not included in the aggregate report produced by the STS. Operations per averaged 4-year data collection cycle.

Figure 3.

The graph documents the annual growth in the STS Congenital Heart Surgery Database by the cumulative number of operations over time. The current number of cumulative total operations in the STS Congenital Heart Surgery Database is 164,240. The aggregate report from the Spring 2010 Harvest of the STS Congenital Heart Surgery Database included 98,628 operations performed in the 4-year period of January 1, 2006 through December 31, 2009, inclusive, submitted from 85 centers from North America, 84 from the United States of America and 1 from Canada. One Japanese center also submits data; however, these Japanese data are not included in the aggregate report produced by the STS. Cumulative operations over time.

Table 1STS Congenital Heart Surgery Database - Counts of Index Operations, Surgery Years 1998 – 2009^{1,2}

Time Period of Analysis = January 1, 1998 – December 31, 2009	All Index Operations in Database	Preoperative Risk Factor of Asplenia = Yes	Preoperative Risk Factor of Polysplenia = Yes	No Asplenia or Polysplenia
STS-EACTS Category 1 - number of Index Operations	21151	55	47	21049
STS-EACTS Category 2 - number of Index Operations	24399	350	132	23921
STS-EACTS Category 3 - number of Index Operations	11308	164	59	11087
STS-EACTS Category 4 - number of Index Operations	17058	536	92	16431
STS-EACTS Category 5 - number of Index Operations	3237	39	31	3167
Overall STS-EACTS Categories 1–5 - number of Index Operations	77153	1144	361	75655
STS-EACTS Category 1 - number of Index Operations with Discharge mortality = Yes	116	3	5	108
STS-EACTS Category 2 - number of Index Operations with Discharge mortality = Yes	415	16	10	387
STS-EACTS Category 3 - number of Index Operations with Discharge mortality = Yes	301	7	1	292
STS-EACTS Category 4 - number of Index Operations with Discharge mortality = Yes	1392	81	13	1285
STS-EACTS Category 5 - number of Index Operations with Discharge mortality = Yes	672	12	5	647
Overall STS-EACTS Categories 1–5 - number of Index Operations with Discharge Mortality = Yes	2896	119	34	2719
STS-EACTS Category 1 - Percentage Discharge Mortality	0.5%	5.5%	10.6%	0.5%
STS-EACTS Category 2 - Percentage Discharge Mortality	1.7%	4.6%	7.6%	1.6%
STS-EACTS Category 3 - Percentage Discharge Mortality	2.7%	4.3%	1.7%	2.6%
STS-EACTS Category 4 - Percentage Discharge Mortality	8.2%	15.1%	14.1%	7.8%
STS-EACTS Category 5 - Percentage Discharge Mortality	20.8%	30.8%	16.1%	20.4%
Overall STS-EACTS Categories 1–5 - Percentage Discharge Mortality	3.8%	10.4%	9.4%	3.6%

¹Excludes operations performed by STS Database Participants with > 15% missing data for the following fields in the STS Congenital Heart Surgery Database: discharge mortality or non-cardiac abnormalities. Also excludes individual records of operations missing data for the following fields: discharge mortality or non-cardiac abnormalities

²Excludes operations performed on patients weighing less than or equal to 2,500 g undergoing PDA ligation as their primary procedure

Table 2STS Congenital Heart Surgery Database - Counts of Index Operations, Surgery Years 1998 – 2009^{1,2}

Diagnosis	All Index Shunt Operations in Database January 1, 1998 – December 31, 2009 ²	All Index Glenn Operations in Database January 1, 1998 – December 31, 2009 ³	All Index Fontan Operations in Database January 1, 1998 – December 31, 2009 ⁴
All Single Ventricle - number of Index Operations ⁵	804	1669	1936
All Single Ventricle except Heterotaxy - number of Index Operations ⁶	656	1468	1696
Single ventricle, Heterotaxia syndrome - number of Index Operations ⁷	148	201	240
All Single Ventricle - number of Index Operations with Discharge mortality = Yes ⁵	59	24	41
All Single Ventricle except Heterotaxy - number of Index Operations with Discharge mortality = Yes ⁶	43	21	31
Single ventricle, Heterotaxia syndrome - number of Index Operations with Discharge mortality = Yes ⁷	16	3	10
All Single Ventricle - Percentage Discharge Mortality ⁵	7.3%	1.4%	2.1%
All Single Ventricle except Heterotaxy - Percentage Discharge Mortality ⁶	6.6%	1.4%	1.8%
Single ventricle, Heterotaxia syndrome - Percentage Discharge Mortality ⁷	10.8%	1.5%	4.2%

¹Excludes operations performed by STS Database Participants with > 15% missing data for the following field in the STS Congenital Heart Surgery Database: discharge mortality. Also excludes individual records of operations missing data for the following field: discharge mortality

²Includes the following primary procedures:

1. Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)
2. Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery)
3. Shunt, Systemic to pulmonary, Other

³Includes the following primary procedures:

1. Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)
2. Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn)
3. Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn)
4. HemiFontan

⁴Includes the following primary procedures:

1. Fontan, Atrio-pulmonary connection
2. Fontan, Atrio-ventricular connection
3. Fontan, TCPC, Lateral tunnel, Fenestrated
4. Fontan, TCPC, Lateral tunnel, Nonfenestrated
5. Fontan, TCPC, External conduit, Fenestrated

6. Fontan, TCPC, External conduit, Nonfenestrated
7. Fontan, Other

⁵Includes the following primary diagnoses:

1. Single ventricle, DILV
2. Single ventricle, DIRV
3. Single ventricle, Mitral atresia
4. Single ventricle, Tricuspid atresia
5. Single ventricle, Unbalanced AV canal
6. Single ventricle, Heterotaxia syndrome
7. Single ventricle, Other

⁶Includes the following primary diagnoses:

1. Single ventricle, DILV
2. Single ventricle, DIRV
3. Single ventricle, Mitral atresia
4. Single ventricle, Tricuspid atresia
5. Single ventricle, Unbalanced AV canal
6. Single ventricle, Other

⁷Includes the following primary diagnosis:

1. Single ventricle, Heterotaxia syndrome