## Transposition of the Great Arteries in the Developing World



### Surgery and Outcomes

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#### ABSTRACT

**BACKGROUND** Little has been published regarding surgery for transposition of the great arteries (TGA) in the developing world.

**OBJECTIVES** This study sought to identify patient characteristics, surgical interventions, institutional characteristics, risk factors for mortality, and outcomes among patients undergoing surgery for TGA in this setting.

**METHODS** Developing world congenital heart surgical programs submitted de-identified data to a novel international collaborative database as part of a quality improvement project. We conducted a retrospective cohort study that included all cases of TGA with intact ventricular septum and TGA with ventricular septal defect performed from 2010 to 2013. Demographic, surgical, and institutional characteristics and their associations with in-hospital mortality were identified.

**RESULTS** There were 778 TGA operations performed at 26 centers, 480 (62%) for TGA with intact ventricular septum and 298 (38%) for TGA with ventricular septal defect. Most (80%) were single-stage arterial switch operations, but 20% were atrial baffling procedures (atrial switch operation) or 2-stage repairs (pulmonary artery band followed by arterial switch operation). Age at operation was >30 days in one-half of the cases and did not vary significantly with operation type. Survival was 85% and did not significantly vary with age at operation or operation type. Preceding septostomy was infrequently reported (16%) and was not associated with surgical mortality. Mortality was associated with lower World Health Organization weight/body mass index-for-age percentile and lower institutional volume of TGA repair.

**CONCLUSIONS** Surgical repair of TGA performed in the developing world is associated with an early survival of 85%. Type of surgical repair and age at operation varied considerably, but no associations with mortality were identified. In contrast, poor nutrition and small surgical volume were most strongly associated with mortality. Multicenter collaborative quality improvement efforts may benefit patients with TGA in the developing world. (J Am Coll Cardiol 2017;69:43-51) © 2017 by the American College of Cardiology Foundation.



Survival and improving quality of life for children with previously lethal cardiac malformations (1). This is important progress, but mortality

associated with CHD and its surgical care remains challenging (2).

D-loop transposition of the great arteries (TGA) is the second most common form of cyanotic congenital disease (3). Left untreated, TGA is associated with

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#### ABBREVIATIONS AND ACRONYMS

ASO = arterial switch operation

ATS = atrial switch BMI = body mass index

CHD = congenital heart disease

IQIC = International Ouality

Improvement Collaborative for Congenital Heart Surgery in Developing World Countries

IVS = intact ventricular septum

**STS** = Society of Thoracic Surgeons

TGA = transposition of the great arteries

VSD = ventricular septal defect

WHO = World Health Organization mortality approaching 85% to 90% (4,5). The medical and surgical management of TGA is well established in many North American and European countries, with the usual practice being single-stage anatomic repair with arterial switch operation (ASO) in the first week of life (3,6,7). Thirty-day mortality is <3%, and 20-year survival approaches 90% (6,8-10). However, less is known about surgery for TGA in the developing world.

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The purpose of this study was to identify patient characteristics, surgical interventions, institutional characteristics, risk factors for mortality, and outcomes among patients with TGA in the developing world, using data from the International Quality Improvement Collaborative for Congenital Heart Surgery in Developing World Countries (IQIC), a novel, multicenter effort aimed at reducing mortality in developing world settings (Figure 1) (11).

#### METHODS

Twenty-six IQIC sites in 15 countries contributed data to this study (Table 1). All sites self-identified as practicing in a resource-limited environment and participated voluntarily in IQIC. Sites submitted deidentified data pertaining to patients <18 years of age undergoing operative repair for CHD that were collected for quality improvement purposes. Data were verified using a random 10% sample of cases from each site. Key variables audited included type of procedure, Risk Adjustment for Congenital Heart Surgery 1 (RACHS-1) category, age, prematurity, post-operative outcomes, and 30-day follow-up outcomes. Approval to conduct research using the IQIC database was obtained from the Boston Children's Hospital Institutional Review Board. The IQIC database was searched for all instances of ASO, atrial switch (ATS), and TGA between January 2010 and December 2013. Associated cardiac lesions were noted.

**INCLUSION/EXCLUSION CRITERIA.** Operations for TGA with intact ventricular septum (TGA/IVS) and TGA with ventricular septal defect (TGA/VSD) were included. Patients entered into the database as having double-outlet right ventricle who underwent ASO with VSD closure were included as TGA/VSD.

Operations in which the underlying anatomy included right- or left-sided obstructive lesions, including coarctation of the aorta, were excluded, as were operations that involved complex intracardiac lesions, such as straddling atrioventricular valves. Operations without data pertaining to mortality also were excluded.

**STATISTICAL ANALYSIS.** Demographic, surgical, and institutional characteristics were summarized with frequencies and percentages. In order to account for the correlation among patients within the same institution, generalized estimating equation models were used to evaluate associations with in-hospital mortality. Risk factors significant at the 0.10 level in univariate analysis were considered for inclusion in a multivariable model; p < 0.05 was required for retention in the final model. Odds ratios and 95% confidence intervals were estimated.

#### RESULTS

There were 778 operations performed for TGA from 2010 to 2013 (**Table 2**). Most surgeries (62%) were for TGA/IVS; the remainder (38%) were for TGA/VSD. The majority of surgeries (70%) were performed in males. Patients in a large proportion of cases (48%) were below the 5th percentile for World Health Organization (WHO) weight/body mass index (BMI)-for-age percentile. Surgeries in patients with identified chromosomal abnormalities, major noncardiac structural abnormalities, and prematurity were uncommon. Preceding septostomy was reported in 16% of cases.

Most surgeries (80%) were single-stage ASO, but Mustard or Senning ATS (13%) or 2-stage pulmonary artery banding followed by ASO (7%) occurred frequently. Only 11% of operations were performed during the first week of life. The remaining 89% of operations were performed later, with a large proportion (51%) performed after 1 month of age. Among those in the latter group, 350 surgeries (45%) were performed between 1 month and 1 year of age, and 44 (6%) were undertaken between 1 and 12 years of age. There was no significant association between patient age and choice of surgical repair.

The average annual institutional volume of TGA operations varied. Most centers (62%) performed fewer than 10 TGA repairs annually. There was no association between surgical volume and choice of surgical repair.

Univariate associations with mortality are given in **Table 3**. The overall mortality rate was 15%. VSD closure as part of the operation, male sex, lower WHO weight/BMI-for-age percentile, weight <3 kg, and prematurity were all factors associated with higher in-hospital mortality. Average annual volumes of TGA repair <10 cases and from 10 to 19 cases were also associated with higher mortality relative to average annual volume  $\geq$ 20 cases.

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