Clinical and Functional Developmental Outcomes in Neonates Undergoing Truncus Arteriosus Repair: A Cohort Study

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Background. **Truncus arteriosus (TA) is an uncommon congenital cardiac lesion that portends an exceedingly poor prognosis if not repaired. The objective of this study was to assess the clinical and developmental outcomes in a prospective cohort of patients who underwent TA repair.**

Methods. **All patients who underwent a TA repair between 1996 and 2012 were included. Follow-up clinical, neurologic, and developmental data were obtained from the Western Canadian Complex Pediatric Therapies Follow-up Program database. Functional developmental outcomes were assessed at 21.1 ± 2.5 months of age with the Adaptive Behavior Assessment System-II, General Adaptive Composite (GAC) score. Survival and outcomes were compared between those with and without chromosomal abnormalities (CA). Survival and freedom from reintervention were assessed by Kaplan-Meier analysis.**

Results. **The study comprised 36 infants (19 male). CA was identified in 13, with 22q11.2 deletion in 10 patients. Patients underwent TA repair at a median age of 10 days; 5 patients underwent concomitant interrupted arch repair. There were 8 deaths, 2 of which occurred in the hospital. The 5-year survival was 79.4%. Survival was similar between those with and without CA. At 5 years, freedom from reoperation was 77.2%. The mean GAC was higher in the patients without CA (93.6 ± 12.8 vs 76.1 ± 13.1, $p = 0.0016$).**

Conclusions. **Patients with surgically repaired TA continue to have significant postoperative mortality. Reoperation and cardiac catheterization are eventualities for a quarter of patients in the first 5 years of life. Functional developmental outcome in patients without CA is good, although it is significantly impaired in those with CA.**

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Truncus arteriosus (TA) (or common arterial trunk) is a rare congenital cardiac lesion, constituting 2% to 4% of all congenital heart lesions [1, 2]. It is characterized by a single arterial trunk arising from the heart, supplying the origins of the coronary arteries, the true pulmonary arteries, and the brachiocephalic vessels. If the condition is not repaired early in life, the prognosis is dismal.

The anatomy of this defect has been understood for many decades, but it was not until 1968 that McGoon and colleagues [2] reported a successful strategy for surgical repair. Although initial surgical repairs were fraught with challenges and troubled by poor outcomes, a TA repair can now be completed with a less than 5% risk of operative mortality [3]. Furthermore, long-term studies have shown excellent actuarial survival of 83% at 15 years, although a need persists for reoperations related to right ventricle to pulmonary artery conduit replacements and truncal valve repair or replacement [4].

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Deletion of chromosome 22q11.2 is present in 30% to 35% of cases of TA [5–7]. Previous work done by the Western Canadian Complex Pediatric Therapies Follow-up Program has demonstrated that neonates affected by deletion 22q11.2 undergoing a variety of complex cardiac surgical procedures have significantly worse neurodevelopmental outcomes than do those without the deletion, although patients with TA were not separately assessed [8]. Although survival is similar between those with and without this chromosomal anomaly (CA) [9], understanding the importance of genetic abnormalities in predicting neurodevelopmental outcomes in TA patients is important for counseling of parents and caregivers.

Neurologic and developmental outcomes in patients with surgically corrected TA are largely unknown, inasmuch as currently published studies have focused only on perioperative and long-term survival. The objective of this study was to assess clinical outcomes, survival, and early childhood neurologic and functional outcomes in a prospectively followed up cohort of patients with repaired TA.

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Patients and Methods

All patients who underwent a TA repair at 6 weeks of age or less at the Stollery Children's Hospital in Edmonton, Alberta, Canada, between 1996 and 2012 were included. The Stollery Children's Hospital is a large surgical referral center for the Western Canadian Children's Heart Network. Follow-up clinical and functional data were obtained from the Western Canadian Complex Pediatric Therapies Follow-up Program. This program maintains a large prospectively collected database including detailed developmental outcomes in all children who had complex neonatal ($\leq$6 weeks of age) surgical procedures at the Stollery Children's Hospital [10]. Details of the methodology of this program and the collection of developmental outcome data from follow-up clinics at the tertiary sites of referral have been previously published [11]. Consents are obtained from the patient's legal guardian. Ethics board approvals for this study from all sites were obtained.

Detailed patient demographic, medical, and surgical information were obtained from the database and complemented with chart review. Long-term follow-up, procedural details, and timing of all repeated cardiac catheterizations, catheter-based interventions, and repeated surgical procedures were obtained. Collected and analyzed variables are given in Table 1. Follow-up care is available for all surviving children. Mortality was ascertained at each of the six follow-up sites with direct contact with the families and attending physicians. Comparisons of demographic, operative, postoperative, neurologic, and functional developmental data between patients with and without chromosomal abnormality are shown in Table 1.

When each child was 18 to 24 months old, a nurse research assistant recorded history of hospitalizations, medication use, and physical measurements. Each child was examined by a pediatrician experienced in developmental follow-up, as previously described [11, 12]. Motor or sensory disability was determined if a child had cerebral palsy [13], visual impairment (corrected visual acuity in the better eye $<$20/60), or binaural/bilateral sensorineural hearing loss greater than 40 dB at any frequency from 250 to 4,000 Hz. Functional outcomes were assessed with a parent-completed questionnaire, the Adaptive Behavior Assessment System-II (ABAS-II), General Adaptive Composite score (GAC) [14]. The ABAS-II assesses the adaptive behaviors of individuals within a community and home context. Assessed skills are grouped into three composite scores: conceptual (communication, functional preacademics, self direction), social (leisure, social), and practical (community use, home living, health and safety, self care). These composite scores are combined with the motor skill score to give the overall GAC. The GAC is a functional measure that compares a person's global adaptive skills with the adaptive skills of others in the same age group from the standardization sample. This standardized outcome measure has an age-based population mean of 100, with a standard deviation (SD) of 15, and 2.27% of the population is expected to have score below 70 (2 SD below the mean).

The socioeconomic status of parents was determined with the Blishen Index (population mean, 43; SD, 13) based on employment of the main wage earner of each family [15].

Statistical Analysis

Continuous variables were tested for the normality of their distribution and are presented as means with standard deviation or medians with interquartile range (IQR) and were analyzed with the t test or the Kruskal-Wallis test. Categoric variables are presented as counts and percentages and were analyzed with Fisher's exact test or the χ^2 test. Actuarial survival and freedom from repeated operation and catheter intervention were assessed by Kaplan-Meier survival analysis, and differences between groups were analyzed with the log-rank test. For outcomes other than mortality, patients were censored at the time of death. All p values are two-sided, and statistical significance was defined as a p value of less than 0.05. All analyses were conducted using Stata (College Station, TX, IC version 11).

Results

The study comprised 36 patients, 19 (52.8%) of whom were male (Table 1). TA was diagnosed prenatally in only 28% of patients, and 21 patients were referred to the Stollery Hospital for surgical procedures from out of the region. Truncus type was specified in all patients: 16 patients (44%) type I truncus, 11 (31%) had type II, 7 (19%) had type III, and 2 (6%) had type IV. Seven patients had associated cardiac lesions, including transposition of the great arteries (1) and interrupted aortic arch (IAA) (5) and right aortic arch (2). Chromosomal abnormalities were identified in 13 patients. Ten were found to have del22q11.2, and the remaining 3 had Down syndrome, Robertsonian translocation of 13 and 14, and level 3 mosaicism, each.

Operative Characteristics

Sixteen patients (44%) were intubated and ventilated preoperatively. The patients were generally well going into operation, with minimally elevated lactate and nearly normal pH on average (Table 1). The patients underwent TA repair at a median (IQR) age of 10 days (7, 13.5), with 11 patients undergoing surgical procedures before 7 days of age. The median (IQR) weight at the time of operation was 3.3 kg (2.7, 3.55), with the smallest infant being operated on at 1.7 kg. Four patients in the cohort were delivered at less than 37 weeks gestation.

The surgical technique was at each surgeon's discretion. Five patients, 4 without CA and 1 with 22q11 deletion, underwent concomitant IAA repair. Twenty-five patients required deep hypothermic circulatory arrest (DHCA) for a median (IQR) time of 24 (10, 33) minutes. This is a similar rate used in other series [16]. Four