Pregnancy in Patients with Congenitally Corrected Transposition of the Great Arteries

Research article
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Abstract
Background: There is scarcity of data on pregnancy in women with congenitally corrected transposition of the great arteries (ccTGA) and what has been published, so far, is based mainly on reported small retrospective series. However, in most of these reports patients with dextro-transposition of the great arteries (d-TGA), who underwent surgical repair in form of atrial switch operation, have been added to the ccTGA group as well.

Methods: The clinical records were collected from the medical database of the Prince Sultan Cardiac Center in Riyadh on eight identified women of childbearing age with ccTGA who were evaluated in the last 15 years (2003–2018).

Results: 5 out of 7 women with ccTGA had good right ventricular (RV) systolic function after successful pregnancy while in 2 women it was mildly decreased after pregnancy. The systemic atrioventricular valve regurgitation (SAVVR) worsened to become severe in 2 out of 7 patients after pregnancy while in 5 patients it did not show any significant changes after pregnancy. Three women had complete heart block, 2 of them required permanent pacemaker implantation before pregnancy but all the three did not show any significant changes in RV systolic function and SAVVR after their pregnancies compared to their echocardiographic findings before pregnancy.

Conclusions: The pregnancy is well tolerated in women with ccTGA. There is no significant effect of pregnancy on the RV systolic function but it may increase the SAVVR in some patients. Women with complete heart block with or without Pacemaker can tolerate pregnancy very well.

Introduction
Congenitally corrected transposition of the great arteries (ccTGA) is a combination of atrophicventricular and ventriculoarterial discordance. It is a rare congenital heart defect (CHD), accounting for less than 1% of all CHD [1]. The right ventricle (RV) supports the systemic circulation in patients with ccTGA and guarded by the tricuspid valve. Most patients with ccTGA will develop RV dysfunction, which can often occur by the third decade of life [2]. The maternal cardiac complications in women with systemic RV includes atrial arrhythmias, deterioration in NYHA functional class and RV failure. These complications occur in about one third of patients with RV dysfunction or arrhythmias prior to pregnancy [3,4].

There is scarcity of data on pregnancy in women with ccTGA and what has been published, so far, is based mainly on reported small retrospective series. However, in most of these reports patients with dextro-transposition of the great arteries...
great arteries (d-TGA), who underwent surgical repair in form of atrial switch operation, have been added to the ccTGA group as well.

The primary aim of this study is to review and to add to the literature the pregnant patients with ccTGA only who were being followed before, during and after their pregnancy at our large tertiary cardiac center in Riyadh, Saudi Arabia.

Methods

Retrospectively, we reviewed the charts of patients with ccTGA who were being followed before, during (3rd trimester) and after (6 to 12 months) pregnancy at Prince Sultan Cardiac Center (PSCC) in Riyadh, Saudi Arabia. The focus of this study was on echocardiographic findings (such as RV systolic function and systemic atrioventricular (tricuspid) valve regurgitation (SAVVR) before, during and after pregnancy. These data were collected from the medical database of the PSCC in Riyadh on seven identified women of childbearing age with ccTGA who were evaluated, in the last 15 years (2003 – 2018). Their medical records were reviewed. All seven women had the diagnosis of ccTGA confirmed by one or more of the following: comprehensive echocardiographic examination, cardiac catheterization and surgical intervention. The echocardiographic findings (RV systolic function and degree of SAVVR) were taken from the finalized echo reports of our center’s database while all the echo recordings were reviewed and the findings confirmed by the first author of this study (AAG) as well. The assessment of the RV systolic function was based on the right ventricular fractional area change (RV FAC) (Figure 1). and the overall visual contractility assessment by reviewing cardiologist (AAG). RV systolic function was defined as normal if RV FAC was 35% and above [5-8].

Statistical analysis

Categorical data were presented in tables according to each case. Continuous variables were reported as mean ± standard deviation. Friedman test was used to compare repeated ordinal data. Collected data were entered and summarized in Microsoft Excel. The IBM SPSS software version 25 (IBM Corp. Released 2017.IBM SPSS Statistics for Windows, Version 25.0. Armonk, NY: IBM Corp.) was used for all statistical analysis. A p-value of < 0.05 was considered as statistically significant.

Results

Seven women with ccTGA with a total of 14 successful pregnancies were collected and reviewed at a single center in Riyadh, Saudi Arabia. One (14.3%) had 4 pregnancies, one (14.3%) with 3, two (28.6%) with 2 each, and three (43%) with 1 successful pregnancies, respectively. Their mean age during the last pregnancy was 26.8 ± 3.2 years. Four women (57%) had concomitant cardiac defects such as atrial septal defect (2/4), ventricular septal defects (1/4), and coarctation of the aorta (1/4). Baseline demographic and echocardiographic characteristics are summarized in Table 1.

Three women (43%) had pulmonary artery banding, one at age 6 and the other two at the age of 19 years. Three women (43%) were diagnosed with complete heart block while 2 of them (29%) required permanent pacemaker implantation before their pregnancies at age 12 and 20 years. Both women did not have any significant changes in RV systolic function and SAVVR after their pregnancies compared to their echocardiographic findings before pregnancy. The woman with complete heart block who refused pacemaker insertion did not have any significant changes in RV systolic function and SAVVR during her pregnancy compared to her echocardiographic findings before pregnancy. She delivered by cesarean section due to failure of progress, but with smooth postoperative course. Her average heart rate during pregnancy and delivery was 50 beats per minute. Also, the RV systolic function and SAVVR did not show any changes after delivery. Changes in RV systolic function were observed before, during and after the recent pregnancy, however, these were not statistically significant (RV systolic function changes, p=0.174). All women were having good RV systolic function (RV FAC > 35%) pre-pregnancy. Three (43%) of them showed a mildly decreased RV systolic function (RV FAC 28, 32 and 34%, respectively) during 3rd trimester (7.3±0.5 months) of their pregnancy while only one patient reverted back to normal RV systolic function (RV FAC 38%) after delivery (9.6 ± 6.9 months).

There were no statistically significant changes with regard to SAVVR in all patients ([SAVVR changes, p=0.097]) throughout the pregnancy and after delivery. Two patients (29%) had mild or no changes before, during and after pregnancy. One woman (14%) had moderate SAVVR before, during and after pregnancy and another one had trivial SAVVR before and during pregnancy which increased to a mild degree after pregnancy. In one patient (14%) the SAVVR increased from trivial before pregnancy to severe during pregnancy but returned back to trivial...
after pregnancy. One patient (14%) had moderate SAVVR before pregnancy which increased to severe during pregnancy and sustained after pregnancy.

**Discussion**

Data on pregnant women with ccTGA is limited and what has been reported, so far, is based mainly on small retrospective series. However, in most of these series patients with d-TGA, who underwent surgical repair in form of atrial switch operation, have been added to the ccTGA group as well.

We have reviewed in this study data from pregnant patients with ccTGA only who were being followed before, during and after their pregnancy at a large tertiary cardiac center to add more information to the limited literature found on outcome of the pregnancies in this specific patient population.

Heidi et al.\(^{(6)}\) reported that in most women with ccTGA, the pregnancy can be successful but the rate of cardiovascular complications and fetal loss are increased. The authors studied 60 pregnancies in 22 women resulting in 50 live births (83%). There were one successful twin pregnancy and 11 unsuccessful pregnancies. One patient developed congestive heart failure late in pregnancy because of SAVVR and required valve replacement in the early postpartum period. One patient had a total of 12 pregnancies, including 1 twin pregnancy and 2 unsuccessful pregnancies. She had multiple pregnancy-related complications, including toxemia, congestive heart failure, endocarditis and myocardial infarction (single coronary artery). No other serious pregnancy-related maternal complications and no pregnancy-related deaths occurred.

Kowalik et al.\(^{(5)}\) presented the pregnancy and long-term cardiovascular outcome in 13 women with ccTGA who had

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### Table 1: Baseline clinical and echocardiographic characteristics of all patients before their recent pregnancy

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age</th>
<th>No of successful pregnancies</th>
<th>Concomitant congenital heart defects</th>
<th>Previous surgery</th>
<th>RV systolic function</th>
<th>SAVVR</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>30</td>
<td>4</td>
<td>-</td>
<td>s/p PA banding at age of 18 yrs s/p PAB tightening at age of 19 yrs</td>
<td>Good</td>
<td>Moderate</td>
</tr>
<tr>
<td>2</td>
<td>24</td>
<td>1</td>
<td>CoA</td>
<td>s/p CoA repair at age of 15 days</td>
<td>Good</td>
<td>Mild</td>
</tr>
<tr>
<td>3</td>
<td>23</td>
<td>2</td>
<td>-</td>
<td>s/p PA banding at age of 6 yrs s/p PPM implantation at 20 yrs</td>
<td>Good</td>
<td>Trivial</td>
</tr>
<tr>
<td>4</td>
<td>25</td>
<td>1</td>
<td>ASD</td>
<td>s/p PA banding at age of 19 yrs</td>
<td>Good</td>
<td>Moderate</td>
</tr>
<tr>
<td>5</td>
<td>27</td>
<td>1</td>
<td>ASD</td>
<td>s/p ASD closure and PPM at age of 12 yrs</td>
<td>Good</td>
<td>Trivial</td>
</tr>
<tr>
<td>6</td>
<td>31</td>
<td>2</td>
<td>VSD, PS</td>
<td>-</td>
<td>Good</td>
<td>Mild</td>
</tr>
<tr>
<td>7</td>
<td>28</td>
<td>3</td>
<td>-</td>
<td>-</td>
<td>Good</td>
<td>Moderate</td>
</tr>
</tbody>
</table>

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**Figure 1:** Right ventricular systolic function changes.

**Figure 2:** Systemic atrioventricular valve regurgitation changes pre-pregnancy to after delivery.
a total of 20 pregnancies. Cardiovascular complications during pregnancy and childbirth occurred in 3 patients (16% of successful pregnancies). Two women developed supraventricular arrhythmias at the end of the second trimester; however, they were observed and required no pharmacological treatment. One patient required premature delivery at 37 weeks for documented deterioration of RV systolic function. Another patient with complete heart block without pacemaker underwent prophylactic temporary pacing during delivery. There were no pregnancy-related maternal deaths.

In the present study, RV systolic function after pregnancy remained same as before pregnancy in 5 out of 7 patients and was mildly decreased after pregnancy in only 2 patients. The SAVVR worsened to become severe after pregnancy in 2 out of 7 patients compared to before pregnancy and did not show any significant changes in the remaining 5 patients. Our data indicates that the pregnancy is well tolerated in patients with ccTGA, however, it may increase SAVVR in some patients. One women had complete heart block without a pacemaker and tolerated the pregnancy and delivery well without any cardiac intervention.

**Study limitations**

This study is a retrospective study and thus subject to referral bias. Since ccTGA is a rare CHD and our study comprising of data from a single center, the number of cases enrolled in this cohort is small. Because cardiac MRI were not done for any women enrolled in this study, the assessment of RV function were done only by echocardiogram.

**Conclusions**

The pregnancy is well tolerated in women with ccTGA. There is no significant effect of pregnancy on the RV systolic function but it may increase the SAVVR in some patients. Women with complete heart block with or without pacemaker can tolerate pregnancy very well.

**Acknowledgments**

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**References**