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CLINICAL ARTICLE

Pregnancy and long-term cardiovascular outcomes in women with congenitally corrected transposition of the great arteries

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ABSTRACT

Objective: To define maternal/neonatal outcomes and long-term cardiovascular effects of pregnancy in women with congenitally corrected transposition of the great arteries (ccTGA). *Methods:* Clinical records of all women with ccTGA who were followed at a tertiary care center in Poland between April 1991 and April 2012 were retrospectively reviewed. *Results:* Of the 20 pregnancies among 13 women identified, 19 (95%) were successful. Of the 19 deliveries, 14 (74%) were vaginal and 5 (26%) were cesarean. Cardiovascular complications during pregnancy and childbirth occurred in 3 patients (16% of successful pregnancies). Two women developed supraventricular arrhythmias; they were observed and required no pharmacologic treatment. One patient required premature delivery for documented deterioration of right ventricular function. There were no pregnancy-related maternal deaths. In 1 case, congenital heart disease was diagnosed in the offspring. With regard to long-term follow-up, no differences were found in terms of heart failure admissions, pharmacologic treatment, deaths, or echocardiographic parameters compared with non-pregnant women with ccTGA. *Conclusion:* Successful pregnancy can be achieved by most women with ccTGA. The most common cardiovascular complications are supraventricular arrhythmias but pregnancy does not seem to impair right ventricular function in the long term. Nevertheless, preconception counseling and tertiary care during pregnancy for women with ccTGA are recommended.

1. Introduction

Among the congenital cardiac defects, there are 2 clinical conditions in which the morphologic right ventricle (RV) sustains the systemic circulation: dextro-transposition of the great arteries (d-TGA) following atrial (physiologic) repair (Senning and Mustard procedures); and levo-transposition of the great arteries (congenitally corrected transposition of the great arteries [ccTGA]) [1]. Congenitally corrected transposition of the great arteries is characterized by atrioventricular and ventriculo-arterial discordance. It is a rare defect, accounting for less than 1% of congenital heart disease [2]. Patients with ccTGA as a lone anomaly do not require surgical repair and may be diagnosed accidentally in adulthood; however, other abnormalities (including ventricular septal defect, pulmonary stenosis, and malformation of the tricuspid valve in the systemic position) often coexist. Most women with ccTGA reach childbearing age. The morphologic RV is not designed to support systemic circulation and is predisposed to dysfunction. Pregnancy-induced changes in the cardiovascular system—including a 50% increase in blood volume, increased cardiac output, reduction in cardiovascular resistance, and rapid fluid shifts during delivery—may lead to right ventricular failure in women with a systemic RV [3].

The aim of the present study was to determine maternal and neonatal outcomes, and long-term cardiovascular effects of pregnancy in women with ccTGA.

2. Materials and methods

Data on all adult female patients with ccTGA followed at the Department of Congenital Heart Diseases at the Institute of Cardiology (a tertiary care center in Warsaw, Poland) between April 1, 1991, and April 30, 2012, were retrospectively reviewed. The study took into account concomitant lesions, cardiac procedures prior to pregnancy, number of pregnancies, and age at booking. A detailed obstetric history was obtained directly from patients, and discharge reports from the obstetrics department were reviewed. The study was approved by the local ethics committee, and all patients gave written informed consent.

Cardiovascular complications (development of heart failure, presence of arrhythmias, occurrence of cerebrovascular events) and maternal cardiac mortality during or after pregnancy were analyzed. The diagnosis of cardiovascular complications was based on physical examination, electrocardiography at rest, transthoracic echocardiography, and 24-hour electrocardiography monitoring.

We assessed the incidence of pregnancy-related disorders (gestational hypertension, pre-eclampsia, eclampsia, and gestational diabetes mellitus) and the mode of delivery (spontaneous/assisted vaginal delivery and cesarean delivery). Number of births plus timing of delivery,

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incidence of small for gestational age (SGA; birth weight below the 10th percentile), incidence of spontaneous abortion, perinatal and neonatal mortality, and occurrence of congenital heart disease in offspring were also noted. Premature birth was defined as delivery before 37 weeks; spontaneous abortion was defined as a spontaneous loss of pregnancy at or before 22 weeks. Gestational age was estimated based on last menstrual period. Birth weight percentiles were calculated using maternal ethnicity, gestational age, and gender of the child [4].

The long-term influence of pregnancy on clinical status and echocardiographic parameters was also analyzed among pregnant and non-pregnant women. To avoid the potential influence of age, the youngest nulliparous women (those aged 21–25 years [n = 6]) were excluded from the comparison. Echocardiographic reports at last review were studied, and right ventricular systolic function (assessed visually by an experienced operator as preserved or mildly, moderately, or severely impaired) and degree of tricuspid (i.e. systemic atrioventricular valve) regurgitation were taken into account.

Between-group frequencies were compared via χ^2 or Fisher exact test. The Mann–Whitney *U* test was used to compare continuous variables. STATISTICA version 10 (StatSoft, Tulsa, OK, USA) was used for all data analysis. $P < 0.05$ was considered to be statistically significant.

3. Results

In total, 20 pregnancies in 13 women were identified among the 26 women with ccTGA who were followed at the study center. None of the women underwent surgery prior to pregnancy. Baseline characteristics of the pregnant women are shown in Table 1. Nineteen (95%) pregnancies were successful and there were no perinatal/neonatal deaths. Details on obstetric/fetal complications are summarized in Table 1.

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 pharmacologic treatment. One patient required premature delivery at 37 weeks for documented deterioration of right ventricular function. Another patient with complete heart block without pacemaker underwent prophylactic temporary pacing during delivery. There were no pregnancy-related maternal deaths.

No pregnancy-related disorders were noted. One patient with type 1 diabetes mellitus was treated with an insulin pump during pregnancy and experienced no complications.

Mean duration of follow-up after pregnancy was 19 ± 15 years. Right ventricular dysfunction and tricuspid regurgitation severity at last review are shown in Table 1.

Evaluation of long-term follow-up showed that the rate of hospitalization for heart failure was similar in pregnant and non-pregnant women, as was the rate of pharmacologic treatment of heart failure (Table 2). Echocardiographic parameters (systemic right ventricular function and tricuspid regurgitation severity) did not differ significantly between the groups (Table 2). There were 2 deaths (1 in each group), both caused by severe right ventricular failure.

4. Discussion

Data on pregnancy in women with ccTGA are scarce and based mainly on reported small retrospective series. Several risk scores developed to predict maternal cardiovascular risk, such as CARPREG and ZAHARA, do not account for the systemic RV—probably because of underrepresentation of women with ccTGA in these studies [5–7]. When analyzing pregnancy outcomes in women with a systemic RV, some authors combined data regarding patients with d-TGA after atrial switch repair and data regarding patients with ccTGA. For example, Jain et al. [8] reported that, among patients with congenital heart disease, women with a systemic RV were more likely to develop cardiovascular complications (heart failure, arrhythmia, stroke, cardiac arrest/death)

Table 1
Baseline characteristics, cardiac and obstetric complications of pregnancy, mode of delivery, and echocardiographic parameters at last review in pregnant women with ccTGA.

Patient number	Number of pregnancies	Age at first pregnancy, y	Concomitant lesions	Cyanosis	Complete heart block	Mode of delivery	Cardiac complications	Obstetric complications	Follow-up, y	TR severity	Right ventricular dysfunction
1	1	28	VSD and pulmonary stenosis			Cesarean (increased cardiovascular risk)	Supraventricular arrhythmias	SGA	2	Mild	Mild
2	1	19			+	Cesarean (obstetric reasons)			20	Moderate	Moderate
3	1	27	Situs inversus		+	Cesarean (increased cardiovascular risk)	Systemic right ventricular dysfunction	Preterm delivery; ASD and PDA in offspring	2	Mild	Moderate
4	1	35				Cesarean (increased cardiovascular risk)			2	Mild	Mild
5	2	21	ASD			Vaginal (forceps)			41	Moderate	Moderate
6	2	22				Vaginal			28	Severe	Mild
7	2	31	VSD and pulmonary stenosis	+		Vaginal (ventouse)		One spontaneous abortion	2	Mild	Mild
8	1	31				Cesarean (obstetric reasons)	Supraventricular arrhythmias		2	Mild	Mild
9	3	31				Vaginal			32	Severe	Severe
10	1	22	VSD, ASD, and pulmonary stenosis			Vaginal (ventouse)			34	Mild	Mild
11	2	35				Vaginal		Two preterm deliveries, SGA (first pregnancy)	27	Mild	Moderate
12	2	20	ASD			Vaginal			34	Severe	Moderate
13	1	24	VSD		+	Vaginal	Prophylactic temporary pacing during delivery		25	Moderate	Mild

Abbreviations: ASD, atrial septal defect; ccTGA, congenitally corrected transposition of the great arteries; PDA, patent ductus arteriosus; SGA, small for gestational age; TR, tricuspid regurgitation; VSD, ventricular septal defect.