Not only life expectancy but also quality of life have improved immensely in patients with congenital heart defects in the last decades due to therapeutic advances. The aim of this study was to examine obstetric and cardiac problems during pregnancy after Mustard/Senning repair for transposition of the great arteries.

Methods and Results: Sixty pregnancies in 34 women from 3 centers were studied. The women were interviewed, and their records reviewed for clinical status and diagnostic evaluation. Age range was 16–34 years during first pregnancy, and all were in a low functional class. There were 11 miscarriages and 5 abortions. Of 44 successful pregnancies, 20 were vaginal deliveries and 24, cesarean sections. A total of 25% were delivered prematurely. Thirteen babies had birth weight <2,500g. Deterioration in functional class occurred in 7 pregnancies, without recovery in 5. Deterioration in systolic function occurred in 4 of 44 echocardiographically documented pregnancies, without recovery in 75%. In 2 women resuscitation was necessary during delivery, in 1, supraventricular tachycardia occurred during labor.

Conclusions: Pregnancy is usually well-tolerated, but outcome is unforeseeable and life-threatening problems can occur. These women belong in cardiac care conducted by experienced congenital cardiologists, who systematically check for typical residua. The pregnancy should be planned and gynecologists/obstetricians with special expertise integrated into the consultations. During delivery a congenital cardiologist, and an anesthetist experienced in congenital cardiology, should be present for possible severe cardiac events. (Circ J. 2014; 78: 443–449)

Key Words: Atrial switch operation; Delivery; Pregnancy; Transposition of the great arteries
whereas women with heart disease can develop substantial problems that endanger mother and child.\(^5\)\(^6\) Therefore, the aims of this study were to assess the impact of pregnancy on the course of TGA, to assess the impact of TGA on the pregnancy and fetus, and to survey the occurrence of cardiac, obstetric and fetal complications.

**Methods**

The databases of 3 tertiary care centers for adults with congenital heart disease (CHD) were scanned for women with TGA after Senning/Mustard repair who had a documented pregnancy. A total of 34 women were identified with 60 pregnancies. The records were reviewed for complete diagnosis, residual complications, clinical status, examination results and pregnancy documentation. Most patients had been examined multiple times before, during and after pregnancy. Because of multiple pregnancies and changing medical status between pregnancies, each pregnancy was considered an independent event.

Cardiac evaluation consisted of past medical history, clinical examination, electrocardiography and at least transthoracic echocardiography. Gynecological-obstetric assessment consisted of anamnestic details about the pregnancy, birth and puerperium, as well as anamnestic and clinical information about the child. The patients were contacted and the birth records—where possible—reviewed. Functional class (FC) was determined by medical history and clinical documentation. Each woman was allocated 1 of 4 FC according to Perloff.\(^7\)

**Results**

**Patients**

Patient age during the first pregnancy was between 16 and 34 years (median, 25 years). Patients had in 20 cases an isolated TGA and in 14 cases complex TGA. All patients were operated on using the Mustard (n=17) or Senning (n=17) technique. The median age at the time of operation was 19 months (range, 3–168 months). Four patients needed a second operation due to pulmonary venous obstruction, baffle leak, coarctation of the aorta or banding of the pulmonary artery. The median follow-up after first pregnancy was 4 years (range, 0.5–17 years).

**FC**

All patients were in a good FC before their first pregnancy: FC I (n=28), FC II (n=5), unknown (n=1). Deterioration in FC occurred in 7 cases, but also improved in 1 woman after her second pregnancy (Figure). In 5 of the patients with deterioration there was no recovery to the original FC. The median interval between deterioration and final documentation was 14 months (range, 6–120 months).

**Echocardiographic Data**

Echocardiographic data, before and after pregnancy, were obtained for 44 pregnancies with a median interval of 11 months before and 6 months after pregnancy (Table 1).

**Cardiac Outcome**

With regard to the women with sinus node dysfunction only 3 required a pacemaker before first pregnancy. In 1 further patient sinus node dysfunction was detected many years after her first pregnancy, without a direct correlation with the pregnancy. Here a pacemaker was also implanted and a β-blocker was required due to supraventricular tachycardia.

Ventricular tachycardia with necessary resuscitation and implantation of an implantable cardioverter defibrillator (ICD) was present in 1 patient 2 years before first pregnancy. This pregnancy was well-tolerated without any medication or cardiac complication. Two women required ICD due to pregnancy and birth-related complications.

The rhythm profiles, pacemaker and ICD data, as well as medication taken before and new medication started during or after pregnancies are listed in Table 2.

Development of complications during pregnancy and delivery was investigated. A detailed description of major cardiac complications (symptomatic tachycardia, heart failure, cardiac arrest) during the course of pregnancy is given in Table 3.
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**Obstetric Results**

Forty-four of the 60 pregnancies were successful. Nineteen women had 1 pregnancy, 8 women had 2, 4 women had 3, 2 women had 4 and 1 woman had 5 pregnancies. There were 11 miscarriages (18.3%) and 5 abortions (8.3%; 3 prophylactic; 1 due to cardiac decompensation; and 1 for social reasons). Spontaneous vaginal delivery occurred in 16 cases (36%), with forceps assistance in a further 4 cases (9%). A cesarean section was conducted in 24 cases (55%). Poor health, dyspnea, hypertrophic baby, position of baby, umbilical cord complications, fetal distress, pre-eclampsia and previous cesarean section, were the reasons indicated. In one-third, the decision was based solely on the mother’s wishes.

During pregnancy various complications occurred, alone or combined: pre-eclampsia, vaginal bleeding, premature contractions/premature rupture of membranes, threatened abortion, and cervical incompetence with premature birth (Table 4).

**Fetal Outcome**

Delivery occurred at a median of 39 weeks, 11 babies (25%) were born prematurely at <37 weeks gestation, and 4 of these (9%) were born at <34 weeks. Thirteen infants (31%) weighed <2,500 g (3 of whom were <1,500 g), and 5 of them were small for gestational age (SGA). The median birth weight was 2,910 g (range, 910–4,160 g). There was no case of CHD in the infants (Table 4).

**Discussion**

The present study is one of the largest series of pregnant
Impairment of ventricular function is clinically perceivable through the deterioration in functional ability. In the current study functional deterioration occurred in 14.8% of patients, most commonly a decrease of 1 FC. The most important residua and sequelae after TGA are right (systemic) ventricular dysfunction, tricuspid valve regurgitation (= systemic atrioventricular [AV] valve), baffle leaks, obstruction of the systemic or pulmonary venous baffle, subpulmonary stenosis and rhythm disorders.

Furthermore, problems may affect the offspring, resulting in miscarriage, preterm birth or CHD.

With increasing age, especially after the third decade, dysfunction of the morphological systemic RV can occur. The causes are multifactorial, including hypoxia, ventricular septal defect and abnormal ventricular geometry. Echocardiographic data show that all heart chambers in-
The subpulmonary LV often tolerates the high afterload without symptoms.

After ASO it cannot be predicted when, or if, a significant functional deterioration of the RV will occur. Furthermore, it is not possible to predict if a following pregnancy will be well-tolerated, even after an uneventful first pregnancy.

After ASO, especially in complex TGA, structural or functional tricuspid valve regurgitation (ie, systemic AV valve) can occur. The occurrence of tricuspid regurgitation due to RV impairment and dilation of the tricuspid valve ring is of utmost importance.15

In the current study there was progression in only 3 of the 40 cases of echocardiographically documented tricuspid valve regurgitation. This is much lower than in the Guédès et al study, in which increased regurgitation occurred in 50% of cases.3

Obstructions of the systemic venous or pulmonary venous baffle can occur at the venoatrial anastomosis site or within the corresponding atrium, caused by the position of the suture line, the extent of the atrial excision, or patch size, form or material. In succession, fibrosis, endothelialization and shrinking can occur. Systemic venous obstructions can be complete or incomplete, and isolated or occur together with a pulmonary venous obstruction. Filling of the ventricle can be obstructed and a fixation of stroke volume can develop.1

In this study there was no progression of a systemic venous obstruction. In 1 case obstruction was newly observed. There was no progression of a pulmonary venous obstruction.

Many TGA patients after ASO have a dynamic or fixed subpulmonary, left ventricular outflow tract obstruction (LVOTO). The subpulmonary LV often tolerates the high afterload without symptoms.

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lower in this series compared to other studies, in which 20–22% of the pregnant women suffered arrhythmias.\textsuperscript{14,17,22–24} This is probably because only symptomatic arrhythmias requiring treatment were considered relevant in the present study.

Routine cardiological and gynecological check-up seems to play an important role in the care of women with ASO. The maternal and fetal risks in the current study were considerable.

An increased rate (up to 18.4%) of hypertensive disorders is described in the literature.\textsuperscript{12,23,25} In this study, however, only 1 woman developed pre-eclampsia.

In the present study, abortions were often conducted prophylactically due to the mothers’ condition, but in 1 case due to pregnancy-related cardiac decompensation. There were 11 cases of spontaneous miscarriage (18.3%), which is higher than in the general population (12–15%).\textsuperscript{26} Drenthen et al reported an even higher miscarriage rate in ASO patients (24.6%).\textsuperscript{14}

Premature birth occurred in 25% of the successful pregnancies, which in almost half of the cases was due to premature onset of labor (16.7% of pregnancies). The cause for premature labor was either obstetric (cervical insufficiency, threatened abortion, vaginal bleeding) or unknown. There were no detectable cardiac risk factors, but studies have shown a significantly higher rate of spontaneous premature onset of labor in women with CHD, which could be due to uteroplacental insufficiency.\textsuperscript{27} The rest of the preterm births were because of cesarean section, either due to cardiac decompensation (18.2%), fetal distress (18.2%), or the mother’s wish for a cesarean section (18.2%), which was then conducted in the 36th week.

In other studies there was an even higher rate of premature birth reported.\textsuperscript{4,6,14,22,28,29} This is higher than in the general population. In Europe the percentage of premature birth <37 weeks of gestation is 6–9%, and, specifically in Germany, 7.5%.\textsuperscript{30,31} Tocolytic therapy or labor induction was required in a few of these patients. There is little information, however, about how well it was tolerated. Further information in this area would be useful, because there can be potential cardiac adverse effects.

It was striking that more than half (n=24, 54.5%) of the deliveries occurred as cesarean sections. From a cardiologicalists’ point of view, a vaginal delivery should be preferred. A cesarean section rarely has an advantage, but usually a higher risk of complications due to the anesthesia, the surgical procedure, and a greater blood loss.\textsuperscript{13,16,24} The fact that one-third of the cesarean sections were conducted due to the mothers’ wish shows the importance of adequate counseling.

In this study there were many serious cardiac complications. Similar cardiac complications have also been described in the literature.\textsuperscript{2,3,13,24,28,29} Given that both pregnancies in which the mother was in FC III before pregnancy had major cardiac complications (Table 3), we advise against pregnancy in this group. Unfortunately, women in FC I before pregnancy also suffered major cardiac events, which shows that all women after ASO need to stay under close cardiological observation during pregnancy and should be monitored continuously during labor, delivery and at least for the first few days afterwards. The fact that most of the issues occurred during delivery should be sufficient reason for a cardiologist, or an anesthetist with experience in cardiology, to be present.

Furthermore, it has been noted in the literature that there is a higher risk of mothers with CHD giving birth to children with CHD. The familial risk in TGA is presumed to be 1.0–1.8%.\textsuperscript{31} In the current study, and in accordance with other studies mentioned in this article, there was no child born with CHD or other congenital disease. Nonetheless, fetal echocardiography is recommended between 18 and 22 weeks of gestation.\textsuperscript{32}

### Study Limitations

This is a retrospective study with no control group, and there may be an additional bias due to the loss of women to follow-up. Without a control group it remains unclear if the changes documented on echocardiography over time are due to the natural course or are pregnancy related. Further research in this field including prospective studies with brain natriuretic peptide data would be useful in identifying patients who are going into cardiac failure.\textsuperscript{33} This study was performed at 3 tertiary care centers for adults with CHD. The prevalence of more complex anomalies in these institutions is likely to be higher than either in community-based hospitals or even in departments for cardiology.

The present data were derived from women living in central Europe (Germany, Switzerland). Generalization of the conclusions to women living in other countries is limited.

### References

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