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Outcome of Pregnancy After the Mustard Operation for Transposition of the Great Arteries With Intact Ventricular Septum

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Objectives. **This study examined the outcome of pregnancy in patients with transposition of the great arteries and an intact ventricular septum after a Mustard operation.**

Background. **Before the introduction of surgical treatment, most children with transposition of the great arteries died in early infancy. A number of these patients have now reached their reproductive years. There is little information about the effect of pregnancy on cardiovascular status, particularly the ability of the right ventricle to adjust to the hemodynamic changes of pregnancy. The outcome for the offspring and their risk of congenital heart disease are also unknown.**

Methods. **Twenty-three female late survivors after the Mustard operation >15 years of age were reviewed in relation to the occurrence of pregnancy and its outcome. Serial echocardiographic estimates of right ventricular volume during pregnancy were made in three local patients.**

Results. **Nine women had 15 pregnancies. They were asymptomatic before pregnancy and remained free from cardiac symptoms during each pregnancy. Right ventricular volume in the three patients studied increased during pregnancy but returned to normal at 8 to 11 weeks postpartum. There were 12 live births, 2 spontaneous abortions and 1 intrauterine death. None of the liveborn infants had evidence of congenital heart disease.**

Conclusions. **In this small group of women with good quality late survival after a Mustard operation, pregnancy was well tolerated. We suspect that the incidence of congenital heart disease in infants of mothers with transposition of the great arteries will be at the lower end of the range for mothers with different types of congenital heart disease, but further data will be needed to confirm this.**

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Before the introduction of surgical treatment most children with transposition of the great arteries and an intact ventricular septum died in early infancy (1). The Mustard operation, introduced in the 1960s (2), and balloon atrial septostomy (3) led to a major improvement in survival (4). The Mustard atrial baffle repair directs pulmonary venous return to the right ventricle and the transposed aorta. Systemic venous blood is directed to the left ventricle.

A number of these patients have now reached their reproductive years. To date there is little information about the effect of pregnancy on maternal cardiovascular status, particularly the ability of the right ventricle to adjust to the hemodynamic changes of pregnancy. The outcome for the offspring and their risk of congenital heart disease are also unknown.

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Methods

Between 1964 and 1983 there were 113 survivors with transposition of the great arteries and an intact ventricular septum after a Mustard type of atrial baffle repair at Green Lane Hospital. The 23 female late survivors >15 years of age were reviewed in relation to the occurrence of pregnancy.

Nine of the 23 women had 15 pregnancies. Four women born between 1965 and 1967 had undergone surgical atrial septectomy (Patients 1 to 4, Table 1) and the five women born after 1967 had been treated with balloon atrial septostomy in the neonatal period (Patients 5 to 9, Table 1). They had undergone the Mustard operation during 1968 to 1970 at 1 to 40 months of age (average 21, median 20). The surgical technique used in these patients has been detailed elsewhere (5).

Cardiac catheterization studies had been undertaken 17 to 31 months (average 24, median 23) postoperatively, when patients were aged 28 to 71 months (average 46, median 39) (6). Severe superior venal caval obstruction was identified in one patient (Patient 4, Table 1) who underwent successful reoperation. Another patient with moderate elevation of pulmonary vascular resistance at the age of 5 years had not shown clinical evidence of progression of pulmonary vascular obstructive disease (Patient 2, Table 1).

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Table 1. Prepregnancy Cardiac Status*

Pt No.	Age at Repair	Age at First Pregnancy (yr)	Baffle Obstruction	Baffle Leak	Tricuspid Regurgitation	Rhythm	Other	NYHA
1	3 y 6 mo	24	–	–	Mild	Sinus	–	I
2	3 y 2 mo	23	–	–	Mild	Sinus	Rpa 6.2 units·m^2, aged 5 yr	I
3	2 y 6 mo	21	Minor SVC	Trivial	Mild	Sinus	–	I
4	2 y 6 mo	24	(Reoperation for SVC)	–	Trivial	Junctional	–	II
5	1 y 8 mo	23	–	–	Mild	Sinus	–	I
6	1 y 4 mo	22	–	Trivial	Not known	Sinus	–	I
7	1 y	19	–	–	Mild	Sinus	–	I
8	8 mo	19	–	–	Trivial	Sinus	Essential hypertension	II
9	1 mo	19	Minor SVC	Trivial	Not known	Junctional	–	II

*Composite data from ongoing reviews. No patient (Pt) had pulmonary stenosis. NYHA = New York Heart Association functional class; Rpa = pulmonary vascular resistance; SVC = superior venal caval obstruction; – = absent.

Serial maternal echocardiography was undertaken in three local patients (Patients 4, 5 and 7, Table 1) at 19 to 22 and 31 to 32 weeks of gestation, and 1 day and 8 to 11 weeks postpartum. Apical four-chamber views with the patient in the left lateral position were used to determine right ventricular volumes and estimation of right ventricular ejection fractions, where volume = 0.849 area^2/length (7,8). The transducer was placed so as to see the tricuspid valve, right ventricular free wall, interventricular septum and apex. The right ventricular endocardium was traced connecting the tricuspid valve at its hinge points by a straight line. The longest length from the midtricuspid annulus to the apex was measured. Frames showing the largest volumes on the cine loop or video replay were taken as end-diastole and those with the least volume as end-systole. At least three nonectopic beats were chosen; measurements were made in triplicate and averaged. Only beats with a well delineated endocardium were selected for analysis. Color Doppler was used to grade tricuspid regurgitation (9).

Blood pressure in pregnancy was categorized according to published criteria (10). Body surface area was estimated according to a standard formula (11).

Results

The prepregnancy cardiac status of the nine women is summarized in Table 1. Of note, none had important baffle obstruction or leak, or significant pulmonary stenosis. Age at onset of the 15 pregnancies ranged from 19 to 26 years (average 23, median 22). None had heart failure or was on cardiac medication. Sinus rhythm was present in seven women; junctional interspersed with sinus rhythm was present in two. Echocardiographic assessment of tricuspid incompetence was trivial in two, mild in five and not documented in two patients.

All nine women remained free from cardiac symptoms during each pregnancy. There were no clinical signs of development of right (systemic) heart failure during pregnancy, labor or puerperium.

Two patients developed transient systemic hypertension and one developed preeclampsia in their first pregnancies. Mild chronic hypertension present in another did not worsen during pregnancy.

Maternal echocardiography. Serial echocardiography was undertaken during pregnancy in three patients. Right ventricular volumes at 19 to 22 weeks of gestation were similar to nonpregnant values obtained in the two patients who had echocardiography 10 to 34 months earlier. At 31 to 32 weeks of gestation, volumes had increased and were maximal 1 day postpartum. Volumes were less 8 to 11 weeks postpartum but were greater than in early pregnancy (Fig. 1, Table 2). There was no significant change in serial ejection fraction calculated from these volumes. All three had echocardiographic evidence of mild tricuspid incompetence that remained similar during and after pregnancy.

Fetal outcome. There were 12 live births, two spontaneous first-trimester abortions and one intrauterine death at 23 weeks of gestation. The mother of this pregnancy had mild essential hypertension not at the level usually associated with fetal demise, and the cause of fetal death remains uncertain. The fetus weighed only 300 g at 23 weeks, well below the expected weight for age. Chromosomes were normal, but no postmortem of the fetus was performed. Fetal echocardiography performed in four surviving infants indicated normal cardiac anatomy.

Live births. In the 12 pregnancies continuing to the third trimester, spontaneous labor occurred in 9 at 35 to 41 weeks, and three were induced at 40 to 41 weeks. Vaginal delivery occurred in 10 infants, assisted by forceps or ventouse extraction in four. Two were delivered by Cesarean section because of fetal indications. All but one were of appropriate weight for gestational age (range 2,170 to 3,869 g). There were six male and six female infants.

There was no clinical evidence of congenital heart disease in the neonatal period or at follow-up, which extended from 6 weeks to 4 years. Examination was performed by pediatricians or pediatric cardiologists in seven infants and all have had later follow-up by family practitioners. One infant with a nonspecific systolic murmur had normal anatomical findings on echocardiography.

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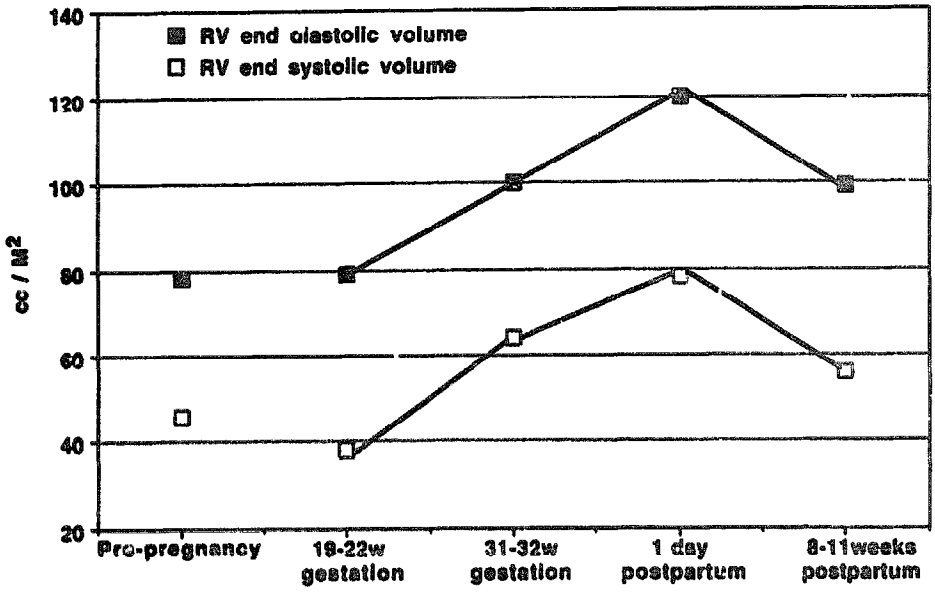

Figure 1. Average values for right ventricular volumes in three patients studied during and after pregnancy.

Discussion

Outcome of pregnancy in transposition of the great arteries. There have been few reports of the outcome of pregnancy in patients with transposition of the great arteries after a Mustard operation in childhood (12–16). Pregnancy outcome in these patients, none of whom appeared to have a large ventricular septal defect, was a minor part of the publication or a single case report. Collectively, they encompass seven pregnancies in six patients. Some appear to have coped well without any obvious hemodynamic deterioration (14–16), but atrial flutter in the third trimester (12) and New York Heart Association functional class deterioration and congestive heart failure (14) were encountered. The infants have been normal (12–16).

One of the late complications of atrial baffle repair for transposition of the great arteries has been right or systemic ventricular myocardial failure (17). The hemodynamic changes of pregnancy, in particular volume loading, are stresses that could induce or exacerbate ventricular failure in such subjects.

None in our small group of patients had clinical signs of hemodynamic deterioration during pregnancy despite the occurrence of transient systemic hypertension in two and preeclampsia in one. It is emphasized that these women represent a group of women late after the Mustard operation with good quality survival, and without any overt cardiac decompensation before pregnancy. The good outcome for pregnancy for this cohort should not necessarily be extrapolated to women who show evidence of right ventricular impairment before pregnancy.

Echocardiography. Right ventricular volumes increased during pregnancy in each, as is the case in pregnancy in normal individuals (18). The values obtained for estimations of right ventricular volumes will depend on the modality and methodology used. The single-plane method of volume determination for the right ventricle has been validated for angiocardiographic measurements (8). We used the echocardiographic apical window, as it is reproducible and we have experience using this method in other adult patients after the Mustard procedure. We found intra- and interobserver variabilities of $r = 0.82$ and $r = 0.70$, respectively, for right ventricular ejection fraction using this method in a group of 24 adults who had had a Mustard operation (19) and a mean right ventricular end-diastolic volume of 102 ± 24 ml/m^2. The first and last observations in our three pregnant patients were well within this range, but measurements had not returned to the initial values at 8 to 11 weeks postpartum. Further observations are required before confident judgements can be made about subtle changes with pregnancy.

Genetic risk. The genetic basis of most congenital heart disease is considered best explained by multifactorial inher-

Table 2. Right Ventricular Volumes

	Pt 4	Pt 5	Pt 7
Prepregnancy			
EDV	82		71
ESV	48		45
19–22 wk			
EDV	91		63
ESV	45		29
31–32 wk			
EDV	132	93	79
ESV	86	55	50
Postpartum 1 day			
EDV	141	122	98
ESV	81	78	69
Postpartum 8–11 wk			
EDV	100	107	98
ESV	52	65	55

EDV = end-diastolic volume (ml/m^2); ESV = end-systolic volume (ml/m^2); Pt = patient.

itance, a type of inheritance in which the genetic predisposition of the individual reacts with the environment to produce the cardiac malformation. The risk of congenital heart disease in offspring of mothers with congenital heart disease is greater than in the general population and varies between 2% and 18% for individual lesions (20). The risk in offspring of mothers with transposition is unknown. There was no evidence of congenital heart disease in the 12 liveborn infants. No mother had any recognized environmental risk factors for congenital heart disease in their offspring. Based on this small cohort and limited comments in the literature, we suspect that the risk of congenital heart disease in offspring will be at the lower end of the range for mothers with different types of congenital heart disease, but further data will be needed to confirm this.

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