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LITERATURE REVIEW

Congenital Heart Defects: Risk Stratification for Pregnancy

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Abstract

Major advances in medicinal and operative therapy on congenital heart defects (CHD) patients have led the majority of affected women survive to childbearing age. The risk of cardiovascular complications during pregnancy and peripartum depends on the type of the underlying defect, the extent and severity of residual haemodynamic lesions and comorbidities. Thuugh pregnancy is well tolerated in patients with CHD, but for some women with particularly high-risk lesions and poor functional class, pregnancy poses significant risk for cardiovascular complications, including premature death. As a result, preconception risk stratification and counseling are mandatory and should be done in all women of childbearing age with CHD. This will enable informed decision making for pregnancy savers.

Keywords: *CHD; risk stratification; pregnancy saver.*

INTRODUCTION

Congenital Heart Defect (CHD) is an abnormality in the structure of the heart or the function of the circulation of the heart that is brought from birth that occurs due to a disturbance or failure of the development of the structure of the heart in the early phase of fetal development. Depending on the magnitude of the hemodynamic dysfunction that occurs, the patient's appearance varies from asymptomatic to very severe.[1]

The incidence of CHD varies between 0.4 – 1.5% of the general population,[1] or about 2800/1,000,000 of the general population.[2] Congenital heart disease occurs in 0.8% of newborn infants around the world.[3,4] Pregnancies obtained by assisted reproductive technology (ART) are associated with an increased risk of complications and congenital anomalies, particularly congenital heart defects (CHDs). The overall prevalence was 1.92%; 1.85% in singletons and 2.23% in twins.[5]

In Indonesia, it is reported that around 45,000 babies are born with CHD every year, and from cases of congenital abnormalities treated at the NICU of RSAB Harapan Kita in 2014 there were 17% of babies with CHD. first week of life, and if not detected early and not treated properly, 50% of deaths will occur in the first month of life.[4] Twenty-five percent of infants with CHD require surgical intervention within the first year of life.[6] Major progress in medical and operative therapy, CHD has led to more than 85% of these infants with CHD surviving to adulthood3 with a better quality of life, and therefore the majority of female sufferers will enter the reproductive phase.1 Women with congenital heart disease (CHD) comprising most patients with heart disease seen during pregnancy, accounting for 80% of all patients.[7]

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Women with CHD are exposed to the risk of cardiovascular complications during pregnancy, which depends on the type of underlying defect, the extent and severity of hemodynamic disturbances, and the comorbidities found.[1] In addition, an important factor that affects the patient's safety during pregnancy, childbirth and the puerperium is the patient's understanding and concern about his or her condition which has more severe risk factors. From a study conducted by Sabanayagam, et al., only 30% of patients with CHD were told that pregnancy was contraindicated for them. Of the 29 women with severe and complex CHD, only 9 (30%) were told that pregnancy was a contraindication for them. For moderate/moderate CHD, only 12 of 39 patients (30.8%) knew that pregnancy was contraindicated for them. [2] It is therefore understandable that the exposure of most women with CHD to pregnancy with the risk of severe cardiovascular complications is inseparable from their ignorance and lack of understanding of the medical conditions they have. This factor of ignorance and misunderstanding can be minimized if every woman with CHD is given pre-conception services, in the form of risk assessment and counseling about pregnancy, childbirth and postpartum in CHD patients with all possible complications. It is hoped that based on the estimated risk, a mature decision will be made, whether to prevent pregnancy or plan a pregnancy. If the choice is to plan a pregnancy, then the pregnancy must be under closer supervision, careful delivery and postpartum planning managed by a multidisciplinary team in order to reduce the risk of morbidity and mortality.

Haemodynamic Changes during Pregnancy

The cardiovascular system needs to adapt during pregnancy and delivery so that the increased hemodynamic requirements can be met.[1] Pregnancy causes hemodynamic changes such as an increase in plasma volume of 40%, an increase in cardiac output and a decrease in systemic vascular resistance. Changes in hemostasis manifest in hypercoagulable conditions.

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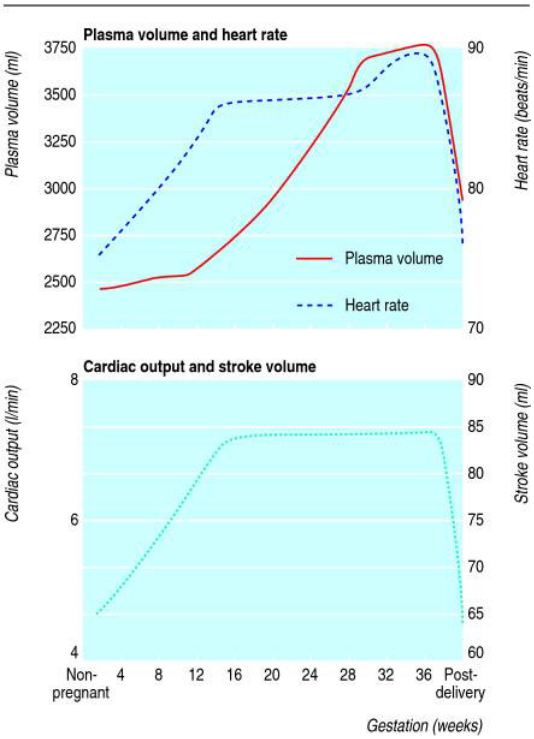

Fig 1 physiological changes of pregnancy[3]

Cardiovascular changes during pregnancy. Plasma volume and cardiac output increase steadily until the end of the second trimester, when cardiac output reaches a plateau at 30-50% above pre-pregnancy levels. Obstructive heart lesions (such as aortic or mitral valve stenosis), which limit cardiac output, are particularly compromised during pregnancy. The increase in blood volume may precipitate heart failure. Cyanosis often worsens during pregnancy as pregnancy related systemic vasodilation may lead to increased right to left shunting.

These physiological changes can be tolerated by women with heart disease, but can also cause complications such as heart failure, arrhythmias and thromboembolic events. The risk for pregnant women with congenital heart disease of having adverse cardiovascular events—such as symptomatic arrhythmia, stroke, pulmonary oedema, overt heart failure, or death—is determined by the ability of their cardiovascular system to adapt to the physiological changes of pregnancy. Different congenital conditions carry specific risks based on their morphological features, previous operations, and current haemodynamic status.[3]

The peripartum period is a very risky condition because during and after delivery of the baby, cardiac demands increase due to uterine contractions, anxiety, straining, autotransfusion due to uterine contractions after delivery of the baby, and blood loss. cardiovascular complications and premature death.[1] All of these risks must be assessed before and during pregnancy.[8]

RISK STRATIFICATION

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The steps of pre-conception efforts consist of risk assessment, risk estimation, informed decision and management planning.[1] The first step according to Rossberg is that before deciding whether a CHD patient will plan a pregnancy (either naturally or with assisted technology), the first question must be The answer is whether pregnancy can take place without significant risk to the mother and fetus. The extent to which the mother is able to cope with the hemodynamic changes resulting from her pregnancy depends on the nature, severity and condition of the underlying heart disease. In this case, the mother is faced with cardiovascular risks and pregnancy risks.[9] Prepregnancy counseling and evaluation is essential in women with CHD, especially those at the highest risk. Unfortunately, most women do not receive appropriate counseling and optimization of their CHD. Without appropriate prepregnancy counseling and optimization of their CHD, women have double the risk of maternal mortality and Heart Failure.[10]

There are three methods proposed in assessing this risk, namely CARPREG, ZAHARA and modified maternal cardiovascular risk classification according to WHO.[11] In this paper, the latter is presented, as follows:

Modified WHO classification of maternal cardiovascular risk

Condition in which pregnancy risk is WHO class I :

- Uncomplicated, minor or mild pulmonary stenosis; patent ductus arteriosus; mitral valve prolapse; successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus and anomalous pulmonary venous drainage); isolated atrial or ventricular ectopic beats.

Condition in which pregnancy risk is WHO classII :

- Atrial or ventricular septal defect not treated by surgery; repaired tetralogy of Fallot; most arrhithmias

Condition in which pregnancy risk is WHO class II-III:

- Mild left ventricular impairment; hypertrophic cardiomyopathy; native or tissue valvular heart disease not considered WHO class I or IV; Marfan syndrome without aortic dilatation; aorta <45 mm in aortic disease associated with bicuspid aortic valve disease

Condition in which pregnancy risk is WHO class III:

- Mechanical valves; systemic right ventricle; Fontan circulation, cyanotic heart disease (unrepaired); other complex congenital heart diseases; aortic dilatation 40-50 mm in aortic disease associated with bicuspid aortic valve

Condition in which pregnancy risk is WHO class IV:

- Pulmonary arterial hypertension of any cause; severe systemic ventricular dysfunction (left ventricular ejection fraction <30%, NYHA class III-IV); previous peripartum cardiomyopathy with any residual impairment of left ventricular function; severe mitral stenosis; severe symptomatic aortic stenosis; Marfan syndrome with dilated aorta >45 mm; aortic dilatation >50 mm in aortic disease associated with bicuspid aortic valve; native severe coarctation.

Risk Class	**Risk of pregnancy**
I	No detectable increased of maternal mortality and no/mild increased in

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	morbidity
II	Small increased risk of maternal mortality or moderate increased in mortality
III	significantly increased of maternal mortality or severe morbidity
IV	Extremely high risk of maternal mortality or severe morbidity; pregnancy contraindicated

Rossberg stated that despite the many morphological and functional variations in heart disease, only a few parameters were associated with complications in pregnancy, namely: a history of previous cardiac events (heart failure, transient ischemic attack or stroke before pregnancy) or arrhythmias; poor functional NYHA class (>II) or cyanosis; LV systolic dysfunction (ejecton fraction <30%); left heart obstruction; and pulmonary artery hypertension).[9]

Pregnant women with CHD are risk factors for the fetus or neonate for spontaneous abortion, premature birth, neonatal events (small for gestational age, respiratory distress syndrome, interventricular hemorrhage, and neonatal death), and high perinatal death.[12] Predict the magnitude of this risk. associated with maternal risk stratification.

Counseling can help people with CHD to realize, understand, and care about their health conditions so that they can play a role in making decisions about themselves. Cauldwell reported the results of a prospective study on pre-conception counseling in patients with CHD that pre-conception counseling; 1) 95% of respondents feel it is an informative service, 2) 95% of respondents feel that gynecologists and cardiologists are very helpful, and 3) 93% of respondents understand the advice given by doctors to them.[13,14]

Issues that must be conveyed in the counseling process must at least contain: [15]

a. Pregnancy risk stratification

1) Maternalcardiac risk

2) Maternalobstetrics risk

3) Fetal and neonatal risks

b. long termeffects of pregnancy on the heart

c. Maternallife expectancy

d. genetic consultation

e. Contraception safety and efficacy

f. Modification of cardiac medications

g. Optimization of cardiac status

h. Planning for pregnancy

Any woman with CHD who has reached puberty should be referred to a cardiac/obstetrics/gynecology clinic/cooperative service for advice on contraception. Likewise

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for women who were first diagnosed with CHD at the reproductive age. Pre-conception efforts on them are focused on counseling, especially aimed at preventing unwanted pregnancies.[16]

For patients who wish to get pregnant, either naturally or with assisted technology, the first thing that must be assessed is the risk stratification. For this reason, it is necessary to carry out careful pre-conception assessment and detailed counseling by experienced cardiologists, obstetricians and other relevant specialists. An important aspect of risk stratification is to decide whether pre-conception interventions, either medication or invasive procedures, can reduce risk. This requires careful consideration because sometimes the action is worse than the disease itself (cure is worse than the disease). The steps for determining cardiovascular risk stratification and fetal complications, counseling and decision making are multidisciplinary and multistep processes, and can be seen in the schematic on the following page.[1]

CONCLUSION

1. Pregnancy in people with CHD exposes them to cardiovascular risks and a higher risk of pregnancy.
2. Risk stratification and pre-conception counseling are efforts to help make more informed decisions so that the desired pregnancy goes more safely