# *Validation of non invasive assessment of pulmonary vascular resistance using Doppler Tissue Imaging of Tricuspid Annulus in Patients with Congenital Heart Diseases*

*Thesis submitted for partial fulfillment of the Master degree*

By

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# ***Introduction***

Congenital cardiovascular defects are present in about 0.8 to 1 percent of live births. They're the most common congenital malformations in newborns. In most cases scientists don't know why they occur. Sometimes a viral infection causes serious problems. German measles (rubella) is an example. If a woman contracts German measles while pregnant, it can interfere with how her baby's heart develops or produce other malformations. Other viral diseases also may cause congenital defects.(1)

Heredity sometimes plays a role in congenital cardiovascular defects. More than one child in a family may have a congenital cardiovascular defect, but this rarely occurs. The risk of having a child with congenital heart disease is higher if a parent or a sibling has a congenital heart defect -- the risk increases from eight in 1000 to 16 in 1000. Certain conditions affecting multiple organs, such as Down's syndrome, can involve the heart, too. Some prescription drugs and over-the-counter medicines, as well as alcohol and "street" drugs, may increase the risk of having a baby with a heart defect.(1)

We can classify congenital heart defects into several categories in order to better understand the problems the baby will experience. They include: (2)

- **Problems that cause too much blood to pass through the lungs** allowing oxygen-rich blood that should be traveling to the body to re-circulate through the lungs, and thus increasing the pulmonary pressure.

- **Problems that cause too little blood to pass through the lungs** allowing blood that has not been to the lungs to travel to the body. The body does not receive enough oxygen with these heart problems, and the baby will be cyanotic.

- **Problems that cause too little blood to travel to the body** as a result of underdeveloped chambers of the heart or blockages in blood vessels that prevent the proper amount of blood from traveling to the body to meet its needs.

In some cases there will be a combination of several heart defects, making for a more complex problem that can fall into several of these categories. (2)

Treatment is based on the severity of the disease. Some mild heart defects do not require any treatment. Others can be treated with medications, percutaneous intervention or surgery. Most adults with congenital heart disease should be monitored by a heart specialist and take precautions to prevent endocarditis throughout their life.(3)

If a patient with a significant congenital heart lesion is not managed early , there will be a chance for pulmonary vascular resistance (PVR) and pulmonary artery pressure (PAP) to increase. This occur as a result of injury to pulmonary vascular endothelium caused by prolonged exposure to increased flow or increased pressure which initiates a cascade of events that involve the release of factors which alter the extracellular matrix , enhance hypertrophy and proliferation of vascular smooth muscle cell and promote connective tissue

synthesis , these changes collectively reduce the diameter of the small pulmonary vessels leading to pulmonary hypertension.(4),(5)

Pulmonary hypertension (PH) refers commonly to elevation of PAP above normal, and is defined as having a pulmonary artery systolic pressure (PASP) of greater than 35mmHg or a mean PAP of greater than 25mmHg.(6)

The development of PH means that PVR will be maintained at a high level. The increased PVR will affect surgical outcome, and surgery is usually contraindicated if PVR is more than half systemic vascular resistance. That is why the assessment of PVR and PAP is a corner stone in management of patients with congenital heart diseases. (7),(8)

Traditionally, PAP & PVR is measured invasively by means of the pulmonary artery catheters (PAC). Although this technique is well established, its invasive nature precludes it from being used in the routine follow up. A noninvasive method of evaluating PVR would (1) allow more frequent assessment of PVR, (2) facilitate the monitoring of individual patient responses, and (3) provide remote-site assessment of PVR.(9)

Because transthoracic echocardiography is an inexpensive, easy, and reproducible method, echo cardiographers continue to search for accurate, noninvasive means of quantifying PVR. Tissue Doppler imaging (TDI) emerged as a new echocardiographic method that can be applied in various clinical conditions.In the year 2007, Gurudevan et al. concluded that Doppler tissue imaging of the lateral tricuspid annulus is a useful clinical tool that can provide a noninvasive estimate of PVR in patients with chronic thromboembolic

pulmonary hypertension. Based on this data, they derived the following logarithmic regression equation:

$$PVR = 3698 - 1227 \times \ln(tSm).$$

Whether or not, these findings can be applied to patients with pulmonary hypertension secondary to congenital heart disease has yet to be proven.

# ***Aim of the study***

The aim of this study is to validate a non invasive method to determine pulmonary vascular resistance in patients with congenital heart disease using Doppler Tissue imaging of the lateral tricuspid annulus, through its correlation with PVR obtained through invasive measurements using cardiac catheterization .

# ***Patients and methods***

Fifty patients with congenital heart disease and pulmonary hypertension (defined as a pulmonary arterial pressure > 25 mmHg at rest [17]) will be enrolled in the study:

Inclusion criteria:

Patients with different congenital heart disease and pulmonary hypertension whether cyanotic or acyanotic.

Exclusion criteria:

- Patients with any contra indication to undergo cardiac catheterization and invasive hemodynamic study.
- Patients with pulmonary stenoses or pulmonary atresia.
- Patients with tricuspid stenoses or tricuspid atresia.
- Patients with truncus arteriosus.

All patients will be subjected to the following:

1- History and clinical examination :

Proper history taking and clinical examination in order to identify the functional capacity of each individual as well as any signs and/or symptoms suggestive of the underlying disease and those suggestive of pulmonary hypertension.

2- 12 lead ECG:

In order to document the presence of any axis deviation, chamber enlargement or hypertrophy, arrhythmia or any other relevant abnormality.

3- Chest X ray:

For the presence or absence of cardiomegaly or any special configuration as well as the pulmonary vasculature.

4- Echocardiography:

Full echocardiographic study using 2D, M mode and color flow Doppler to verify the underlying disease of each individual included in the study will be done within 1 hour of cardiac catheterization.

This will be followed by Doppler tissue imaging (DTI) of the lateral tricuspid valve annulus in the apical four chamber view.

PVR will be measured non invasively using the peak systolic velocity of the tricuspid annulus ( tSm ) using the following equation:

$$\mathbf{PVR = 3698 - 1227 \times In ( tSm )}$$

5- Cardiac catheterization:

All individuals enrolled in the study will be subjected to invasive hemodynamic study by an experienced independent operator unaware of the data obtained from the echocardiographic examination. Invasive measurements of the pulmonary vascular resistance will be done using Fick's principle.

Statistical analysis:

The values of PVR obtained by Tissue imaging of lateral Tricuspid annulus will be applied to the following equation [ $PVR = 3698 - 1227 \times In ( tSm )$] & will be correlated and compared with invasive PVR measurement using statistical analysis .

# ***References***


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assessment of PVR and PAP is a corner stone in management of patients with congenital heart diseases[3,4].

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A noninvasive method of evaluating PVR would (1) allow more frequent assessment of PVR, (2) facilitate the monitoring of individual patient responses, and (3) provide remote-site assessment of PVR[5].

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PVR = 3698 - 1227 x ln(tSm).

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# AIM OF THE STUDY

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# Chapter (1)

# SCALE OF THE PROBLEM

## Incidence of CHD

The incidence of congenital heart disease (CHD) is approximately 8 per 1000 livebirths[7,8]. Many infants who are born alive with cardiac defects have anomalies that do not represent a threat to life, at least during infancy. Almost one-third of those infants, or 2.6 per 1000 live births, however, have critical disease, which is defined as a malformation severe enough to result in cardiac catheterization, cardiac surgery, or death within the first year of life[9]. Today, with early detection and proper management, the majority of infants with critical disease can be expected to survive the first year of life[9].

Estimates of the incidence of specific lesions vary, depending on whether the data are drawn from infants or older children and whether the diagnosis is based on clinical, echocardiographic, catheterization, surgical, or postmortem studies[7-10]. The incidence in other countries is remarkably similar to that reported for the United States[11,12]. Despite these differences in case material, except for bicuspid aortic valve and mitral valve prolapse, it is apparent that ventricular septal defect (VSD) is the most common malformation, occurring in 28% of all patients with CHD (Table 1).

**Table (1):** Incidence of Specific Congenital Heart Defects

| Defect | %age of Cases[a] (Averaged) |
|---|---|
| Ventricular septal defect (VSD) | 28.3 |
| Pulmonary stenosis | 9.5 |
| Patent ductus arteriosus | 8.7 |
| VSD with pulmonary stenosis[b] | 6.8 |
| Atrial septal defect, secundum | 6.7 |
| Aortic stenosis | 4.5 |
| Coarctation of aorta | 4.2 |
| Atrioventricular canal[c] | 3.5 |
| Transposition of the great arteries | 3.4 |
| Aortic atresia | 2.4 |
| Truncus arteriosus | 1.6 |
| Tricuspid atresia | 1.2 |
| Anomalous pulmonary venous connection | 1.1 |
| Double-outlet right ventricle | 0.8 |
| Pulmonary atresia without VSD | 0.3 |

[a]Total number of cases = 103,590. [b]Includes tetralogy of Fallot. [c]Includes partial and complete. SOURCE: Data from references 7–9,11, and 12.

## Gender:

Among 2251 infants with critical CHD in the New England Regional Infant Cardiac Program,[9] 53.7 % were male. Certain defects, however, are more common in one sex than in the other. Aortic stenosis occurs more often in boys (4:1), and

atrial septal defects (ASDs) occur more frequently in girls (2.5:1).

## Complications of CHD

**Table (2):** Complications of CHD in Children

| | |
|---|---|
| Congestive heart failure | Growth retardation |
| Hypoxemia | Pulmonary vascular disease |

### *1- Congestive Heart Failure*

Heart failure is a potentially lethal complication of CHD and occurs in more than 80 % of infants who have malformations severe enough to require cardiac catheterization or surgery within the first year of life[13]. The onset is usually within the first 6 months of life; it is rarely found after 1 year of age without a serious intercurrent problem such as infective endocarditis, pneumonia, or anemia [13].

### *2- Hypoxemia*

*Cyanosis,* a bluish tinge to the color of the skin caused by the presence of at least 3 to 5 g/dL of reduced hemoglobin, is frequently the initial sign of CHD in an infant. It may also be an early sign of pulmonary, central nervous system, metabolic disease or methemoglobinemia. Prompt distinction between cardiac and noncardiac cyanosis, usually by echocardiography, is extremely important, as palliation with $PGE_1$ infusion