Comparative Study between 3D Echocardiography Right Ventricular -Volumes and Functions- and Invasive RV Quantification in Children with Valvular Pulmonary Stenosis Pre and Post Balloon Dilation

Thesis

Submitted in Partial Fulfillment of
Master Degree in ***Cardiology***

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2020

بسم الله الرحمن الرحيم

قالوا

سبحانك لا علم لنا

إلا ما علمتنا إنك أنت

العليم الحكيم

صدق الله العظيم

سورة البقرة الآية: ٣٢

Acknowledgments

The vision for this thesis could only become a reality today because of the kind help and support of many; I would like to take the opportunity to extend my gratitude to them all.

First and foremost, all praise is to ALLAH Almighty for his daily blessings, beyond count or description, and beyond all attempts of thanks and gratitude.

For their relentless and continuous guidance, I am grateful to my supervisors. Their support and luminous feedback provided the backbone of this thesis.

I wish to express my deepest gratitude to ***Prof. Dr. Hebattallah Mohamed Attia****, Assistant Professor of Cardiology, Faculty of Medicine, Ain Shams University, for her invaluable mentorship and for her unremitting leadership, evident at every step of this work. It has been an honor to complete this thesis under her direction.*

I am deeply indebted to ***Dr. Yasmin Abdel Razek Esmail****, Lecturer of Cardiology, Faculty of Medicine, Ain Shams University, for his priceless scientific assistance and the invaluable effort he provided during his supervision of this work.*

Foremost, I would like to express my sincere gratitude to ***Dr. Heba Mohamed Nossier****, for the continuous support of my study and research, for her patience, motivation, enthusiasm, and immense knowledge. Her guidance helped me in all the time of research and writing of this thesis. I could not have imagined having a better advisor and mentor for my study.*

I am delighted to have had ***Dr. Mohamed Rashad****, Lecturer of Cardiology, by my side during this endeavor, both as a mentor and as a friend.*

My heartiest gratitude goes, of course, to my father and mother, always paving the path before me, never ceasing to believe in me. My past and my future are shaped by the warmth of your support.

My thanks also go to my brothers and sister, my friends and my colleagues for their continuous support throughout my life.

Finally, I humbly extend my thanks to the patients who participated in this research, my department and to the great institution of Ain Shams University.

Mostafa Mohamed AbdelMonem Hafez

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List of Abbreviations

Abb.	Full term
2D	*Two-dimensional*
3D	*Three-dimensional*
Angio	*Angiography*
BPV	*Balloon pulmonary valvuloplasty*
BSA	*Body surface area*
ECG	*Electrocardiogram*
EDV	*End diastolic volume*
EF	*Ejection fraction*
ESV	*End systolic volume*
FAC	*Fractional area change*
MPA	*Main pulmonary artery*
MSA	*Membrane stabilizing activity*
PA	*Pulmonary artery*
PLAX	*Parasternal long axis*
PR	*Pulmonary regurgitation*
PS	*Pulmonary stenosis*
PSAX	*Parasternal short-axis*
PV	*Pulmonary valve*
RBBB	*Right bundle branch block*
RPA	*Right pulmonary artery*
RV	*Right ventricle*
RVH	*Right ventricle hypertrophy*
RVOT	*Right ventricular outflow tract*
RVOTO	*Right ventricular outflow tract obstruction*
SD	*Standard deviations*
TAPSE	*Tricuspid annular plane systolic excursion*

INTRODUCTION

Pulmonary stenosis (PS) is a common congenital heart disease. It accounts for approximately 8 - 12% of all congenital cardiac defects ***(Yang and Yi, 2005)***. With an incidence of about 1 per 2000 live births worldwide. In Egypt, its prevalence is 2.3 per 10 000 school children ***(Bassili et al., 2000).***

As an isolated defect, PS is the second most common congenital cardiac defect after VSD.

Patient`s presentation may be as asymptomatic with an incidental murmur if mild PS. With severe PS advanced right ventricular dysfunction and failure occur. Even may present with deep cyanosis and acidosis due to critical PS ***(Mitchell and Mhlongo, 2018).***

The traditional treatment for pulmonary valve stenosis prior to 1982 was surgical valvotomy. The relief of pulmonary valve stenosis by balloon dilatation during cardiac catheterization was first reported in 1982 ***(Kan et al., 1982).***

Percutaneous cardiac catheterization is increasingly used both diagnostically and therapeutically in neonates; in some cases (e.g. critical pulmonary stenosis), it is the first-line therapy ***(Rao et al., 2007)***.

In neonates, less invasive interventions are preferred because open heart surgery cannot be performed at all centers and is associated with a high mortality risk, especially in developing countries. Cardiac catheterization and angiography have transformed the care of children with CHD and have greatly increased the safety and efficacy of surgery for CHD ***(Jenab et al., 2013; Dalla et al., 2015)***. It is one of the classic ways of determining the severity of pulmonary stenosis. Pressure gradients can be measured directly and angiography represents anatomy accurately. Nowadays, cardiac catheterization is used for invasive treatment as well as it is thought that it is a valid method to assess RV volume and function in correlation with 3D Echocardiography.

The Right Ventricle has thin wall, the free wall measuring 2–5 mm, and its muscle mass is one sixth the LV one. It is a crescent-shaped chamber with a high capacitance and a greater ability to handle changes in preload than in afterload. When chronically exposed to increased afterload, the RV can adapt with myocardial hypertrophy, since increase in wall pressure leads to increase in wall stress that, by way of Laplace’s law, can be tempered by increased wall thickness. However, maladaptive changes can occur subsequently that lead to RV dilation and a decreased contractility ***(Alexis et al., 2015)***.

Recently it is well known that the right ventricle (RV) plays an important role in the morbidity and mortality of patient affected by several cardiac and pulmonary diseases. Its function, indeed, can be impaired in a lot of cardiac diseases, such as pulmonary hypertension (PH), congenital heart disease (CHD). Recently, several studies have pointed out the prognostic value of RV function in cardiovascular diseases like HF, in which RV systolic function is one of the major mortality predictors. Ultimately, RV remodeling and RV dysfunction have been associated with a poor prognosis ***(Ozben et al., 2015; Focardi et al., 2015)***.

Three-dimensional echocardiography (3DE) allows us to measure RV end-diastolic volume and ejection fraction (3D-RVEF) irrespective of its shape and more reliably compare it to 2D echocardiogram and evaluate the morphologic and functional remodeling of the right ventricle in congenital heart diseases with pressure overload on the RV such as pulmonary valve stenosis. Accordingly, Echocardiography plays a major role in the assessment and management of pulmonary valve stenosis. It is useful in detecting the site of the stenosis, quantifying severity, determining the cause of the stenosis, and is essential in determining an appropriate management strategy. Ancillary findings with pulmonary stenosis such as right ventricular hypertrophy may also be detected and assessed ***(Simpson et al., 2017)***.

So, there is an importance to correlate between 3D echocardiographic assessment of RV volumes and functions with invasive RV quantification by angiography in children with mainly valvular pulmonary stenosis undergoing balloon pulmonary valvuloplasty, hence the rationale of this study.

Aim of the Work

To correlate between 3D echocardiographic assessment of RV volumes and functions with invasive RV quantification by angiography in children with mainly valvular pulmonary stenosis undergoing balloon pulmonary valvuloplasty pre & post balloon dilation, the immediate impact of relieving obstruction on RV volumes.

Chapter 1

PULMONARY STENOSIS

The normal pulmonary valve is enclosed in a proximal sleeve of free-standing right ventricular infundibulum supporting the fibroelastic walls of the pulmonary sinuses at the anatomic ventriculo-arterial junction. The valvular leaflets are attached in a semilunar fashion across this junction, delimiting the extent of the valvular sinuses ***(Mitchell and Mhlongo, 2018).***

As far as stenosis within the pulmonary outflow tract is concerned, obstruction at the valvular level is by far the most common lesion ***(Fitzgerald and Lim, 2011).***

The fusion along the zones of opposition is usually uniform. It begins peripherally, so that the valvular orifice is narrowed to a central opening. The more the fusion extends towards the centre of the valve, the narrower will be the central opening, and the more severe will be the valvular stenosis ***(Fuster et al., 2009).***