Registry of Patients with Pulmonary Hypertension Presented to Ain-Shams University Hospitals

Thesis

Submitted for Partial Fulfillment of Doctorate Degree in Cardiology

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2019

سورة البقرة الآية: ٣٢

Acknowledgment

First and foremost, I feel always indebted to **ALLAH**, *the Most Kind and Most Merciful.*

I'd like to express my respectful thanks and profound gratitude to **Prof. Maiy Hamdy Elsayed,** *Professor of Cardiology, Faculty of Medicine-Ain Shams University for her keen guidance, kind supervision, valuable advice and continuous encouragement, which made possible the completion of this work.*

I am also delighted to express my deepest gratitude and thanks to **Dr. Hebatallah Mohamed Attia,** *Assistant Professor of Cardiology, Faculty of Medicine, Ain Shams University, for her kind care, continuous supervision, valuable instructions, constant help and great assistance throughout this work.*

I am deeply thankful to **Dr. Dina Adel Ezzeldin,** *Lecturer of Cardiology, Faculty of Medicine, Ain Shams University, for her great help, active participation and guidance.*

I would like to express my hearty thanks to all my family for their support till this work was completed.

Last but not least my sincere thanks and appreciation to all patients participated in this study.

Abdelrahman Elsayed Attia Abdelwahed

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List of Abbreviations

Abb.	Full term
6MWD	*6-minute walking distance*
ASD	*Atrial septal defect*
BNP	*Brain natriuretic peptide*
CBC	*Complete blood count*
CCBs	*Calcium channel blockers*
CHD	*Congenital heart disease*
CI	*Cardiac index*
CMR	*Cardiac magnetic resonance*
CT	*Computed tomography*
DVT	*Deep vein thrombosis*
ECG	*Echocardiography*
EMA	*European Medicines Agency*
ERAs	*Endothelin receptor antagonists*
ES	*Eisenmenger's syndrome*
ET	*Endothelin*
FAC	*Fractional area change*
FDA	*Food and Drug Administration*
GE	*General Electric*
IPAH	*Idiopathic pulmonary arterial hypertension*
IUGR	*Intrauterine growth retardation*
MPAP	*Mean PA Pressure*
NT-proBNP	*N-terminal pro-brain natriuretic peptide*
PADP	*PA Diastolic Pressure*

List of Abbreviations (Cont...)

Abb.	Full term
PAH	*Pulmonary arterial hypertension*
PAPUCO	*Pan African Pulmonary hypertension Cohort*
PDA	*Patent ductus arteriosus*
PDE-5i	*Phosphodiesterase type 5 inhibitors*
PH	*Pulmonary hypertension*
pred.	*Predicted*
PROs	*Patient-reported outcomes*
PVOD	*Pulmonary vascular occlusive disease*
PVR	*Pulmonary vascular resistance*
QoL	*Quality-of-life*
RA	*Right atrium*
RAP	*Right atrial pressure*
RHD	*Rheumatic heart disease*
RV	*Right ventricle*
RVH	*RV hypertrophy*
RVSP	*Right ventricle systolic pressure*
SvO2	*Mixed venous oxygen saturation*
TAPSE	*Tricuspid annular plane systolic excursion*
VE/VCO2	*Ventilator equivalents for carbon dioxide*
VO2	*Oxygen consumption*
VSD	*Ventricular septal defect*
WHO	*World Health Organization*

INTRODUCTION

Pulmonary hypertension is a Patho-physiological disorder that involves multiple clinical conditions and can complicate many cardiovascular and respiratory diseases ***(Sliwa et al., 2012)***.

Registries of patients with pulmonary arterial hypertension (PAH) have been instrumental in characterizing the presentation and history of the disease providing a basis for prognostication. Since initial accumulation of data conducted in the 1980s, subsequent registry databases have yielded information about the demographic factors, treatment, and survival and have permitted comparisons between populations in different eras and environments. Inclusion of patients with all subtypes of PAH has also allowed comparisons of these subpopulations ***(Mocumbi et al., 2011; Thienemann et al., 2016)***.

The epidemiology of pulmonary hypertension and its burden has not been studied yet in Egypt, in 2016, chest hospital in Abbasiya tried to retrospectively study 52 patients with pulmonary hypertension with trial to increase awareness about pulmonary hypertension among its physicians ***(Farrag et al., 2016)***.

Another trial in 2016 studied pulmonary hypertension in adult Egyptian patients with b-thalassemia major and its correlation with natural anticoagulant levels ***(Elbedewy et al., 2015)***.

Aim of the Work

To establish a registry for patients with pulmonary hypertension including their clinical data, echocardiographic evaluation, any intervention and the clinical outcome for patients presented to congenital and structural heart disease unit at Ain shams university hospitals.

Chapter 1
PULMONARY HYPERTENSION

Definition: Pulmonary hypertension is a complex group of disorders which result from different pathophysiologic mechanisms but are all defined by a mean pulmonary arterial pressure of 25 mm Hg or more measured by RHC ***(Galie et al., 2016)***.

PAH–CHD represents a preventable form of PAH in the recent decades, advances in diagnostic procedures and cardiac surgery have resulted in the prevention of PAH in most children with CHD and systemic-pulmonary shunts in Western countries; this is, unfortunately, not yet the case in developing countries ***(Chessa et al., 2017)***.

The clinical classification of pulmonary hypertension according to international pulmonary hypertension guidelines *(Simonneau et al., 2013):*

1- Pulmonary arterial hypertension.

2- Pulmonary hypertension due to left heart disease.

3- Pulmonary hypertension due to lung diseases and/or hypoxia.

4- Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions.

5- Pulmonary hypertension due to unclear and/or multifactorial mechanism.

Pathophysiology:

All congenital heart defects, in which a large intra or extra cardiac communication allows unrestricted pressure and volume overload of the pulmonary circulation, may lead to the development of pulmonary arterial hypertension (PAH), unless repair occurs in early childhood ***(Dimopoulos et al., 2014).***

Pulmonary arterial hypertension (PAH, group 1) is a clinical condition characterized by the presence of pre-capillary PH and pulmonary vascular resistance >3 Wood units, in the absence of other causes of pre-capillary PH ***(Galie et al., 2016)***.

Clinical classification of pulmonary arterial hypertension associated with congenital heart disease *(Rubin et al., 2013; D'alto et al., 2015; Galie et al., 2016)*:

1. Eisenmenger's syndrome (ES):

Includes all large intra- and extra-cardiac defects which begin as systemic-to-pulmonary shunts and progress with time to severe elevation of PVR and to reversal or bidirectional shunting; cyanosis, secondary erythrocytosis, and multiple organ involvement are usually present.

2. PAH associated with prevalent systemic-to-pulmonary shunts:

- Correctable.
- Non-correctable.

Includes moderate to large defects; PVR is mildly to moderately increased, systemic-to-pulmonary shunting is still prevalent, whereas cyanosis at rest is not a feature.

3. PAH with small/coincidental defects:

Marked elevation in PVR in the presence of small cardiac defects (usually ventricular septal defects <1 cm and atrial septal defects < 2 cm of effective diameter assessed by echo), which themselves do not account for the development of elevated PVR; the clinical picture is very similar to idiopathic PAH. Closing the defects is contra-indicated.

4. PAH after defect correction:

Congenital heart disease is repaired, but PAH either persists immediately after correction or recurs/develops months or years after correction in the absence of significant postoperative hemodynamic lesions.

Congenital heart disease accounts for nearly one-third of all major congenital anomalies, nearly 1 in 100 children are born with congenital heart disease (6–10/1000 live births),