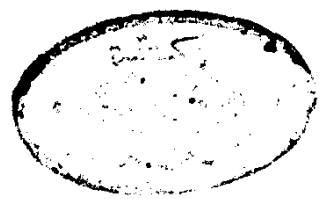
THE MITRAL VALVE IN PATIENTS WITH SECUNDUM ATRIAL SEPTAL DEFECT

THESIS

Submitted in Partial Fulfilment for
The Master Degree of
Cardiology



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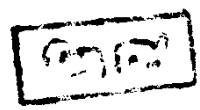

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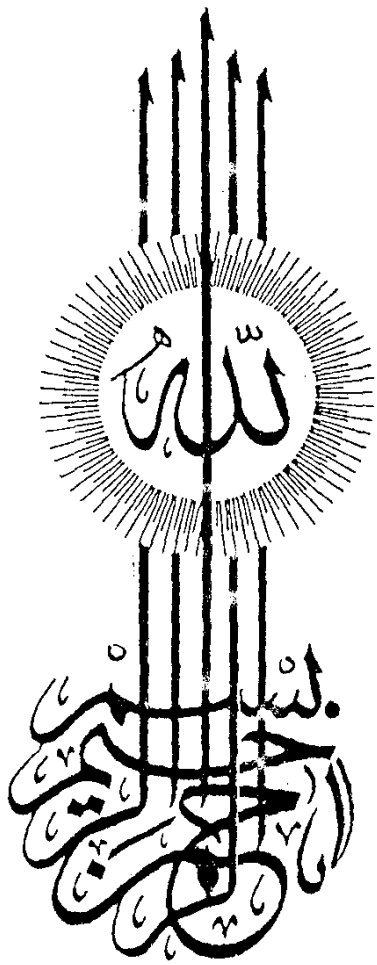

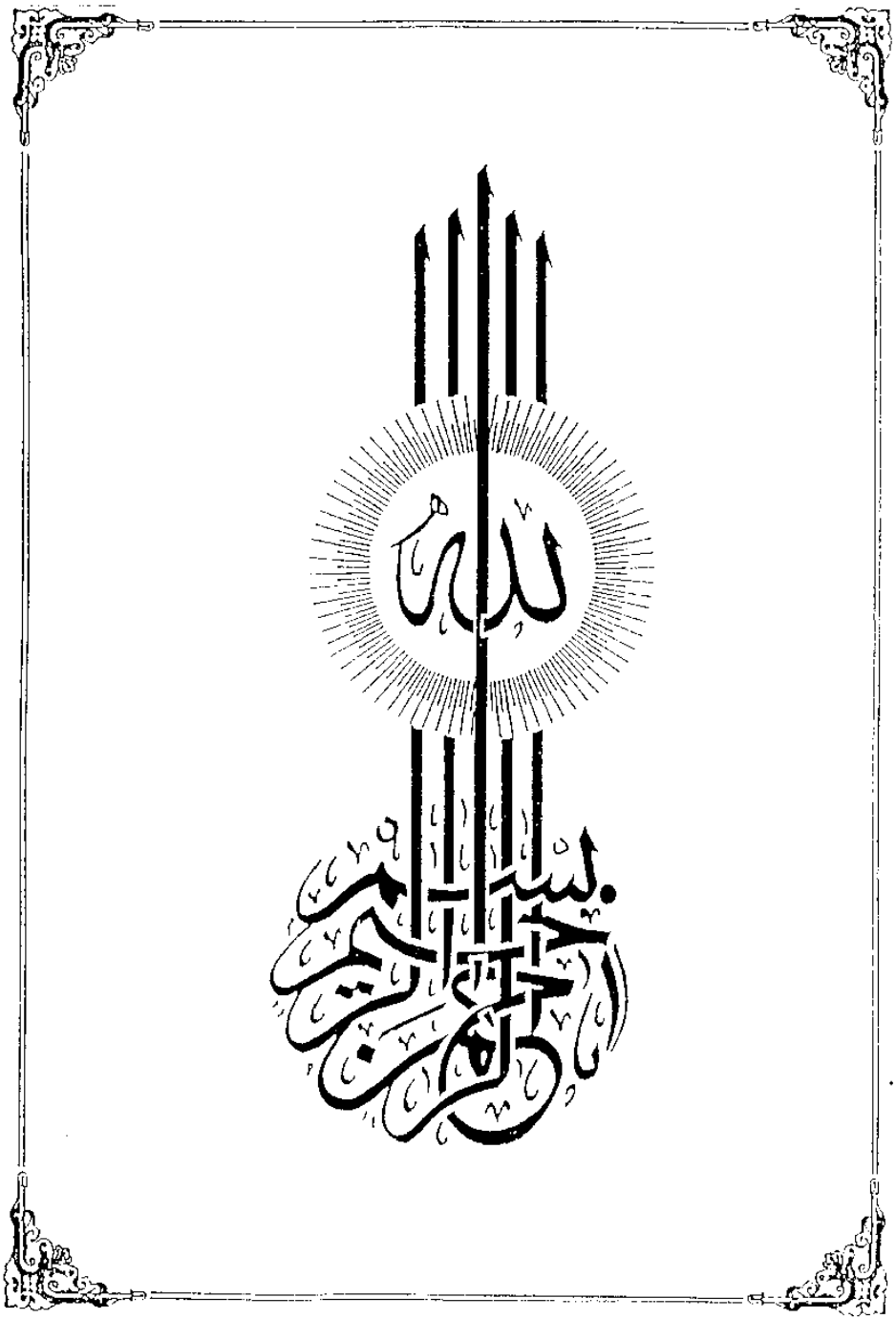

" TO THE MEMORY OF MY FATHER "

C O N T E N T S

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A C K N O W L E D G E M E N T

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INTRODUCTION
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Atrial septal defect was described by Rokitansky in 1875, but it was not until 1941 that Bedford, Papp and Parkinson drew attention to its clinical features (Perloff, 1978).

.. Atrial septal defect (ASD) is a relatively common cardiac malformation. It is found approximately in ten percent of children surviving beyond the first year of life with congenital heart disease (Nadas, 1972). It is the most common form of congenital heart disease among the adults if one excludes the congenital bicuspid aortic valve (Keith, 1978).

An atrial septal defect is a through-and-through communication between the atria at the septal level. The condition is to be distinguished from the valvular-competant foramen ovale (Hurst, 1982).

.. In 1916 Lutembacher published the first comprehensive account of atrial septal defect with mitral stenosis (M.S.), although individual reports had appeared since 1865. (Perloff, 1978).

.. It has been estimated that the incidence of mitral stenosis among patients with ASD is 4 %, while among cases of mitral stenosis, the incidence of atrial septal

defect is 0.6 to 0.7 %. (Steinbrunn et al., 1970).

•• The interatrial septal defect may be of the congenital secundum type or could possibly be an acquired lesion caused by stretching of the margins of the foramen ovale by progressive dilation of the left atrium in mitral stenosis (Goldfarb, 1966).

•• Some patients with Lutembacher syndrome give a history of rheumatic fever, but many do not. Graig and Selzer (1968) pointed out that combination represented a true susceptibility of patients with secundum defects to rheumatic fever, because the frequency of the syndrome was far greater than would occur from random association.

•• Mitral regurgitation combined with secundum atrial septal defect was reported only twice in a series of 100 patients studied by Welch and Associates in 1966. Hynes and Associates (1974), reported mitral regurge in 6 % (46 patients) among all patients with secundum atrial septal defects operated upon in 13 years period at Mayo Clinic. Out of these 46 cases; 6 had rheumatic valvulitis, 8 had redundant mitral tissue, 5 with prolapse, 9 had two or more causes and in 18 patients no cause could be determined.

Leachman and Associates (1976) found angiographic evidence of severe mitral valve prolapse in 16 out of 92 patients with secundum atrial septal defects studied

between 1970 and 1974.

•• The aim of this work is to study the effect of the mitral valve lesions on the clinical aspects and haemodynamics of the associating secundum atrial septal defects.

In the following chapters, the isolated secundum atrial septal defect will be discussed first, then follows a review of literature on the association of mitral valve lesions and secundum atrial septal defect.

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REVIEW OF LITERATURE

PATHOLOGY OF ATRIAL SEPTAL DEFECT

Atrial septal defect is the commonest congenital heart malformation found in adults, but is the fifth or sixth in numerical importance in infancy and childhood. Untreated, death in right-sided heart failure usually occurs at between 30-40 years of age, although survival into the ninth decade was reported. (Pomerance and Davies, 1975).

Bedford (1960), in his study of 185 confirmed cases, found that the vast majority of defects were of the ostium secundum type, with 7 % of the ostium primum type, and 5 % of the sinus venosus type.

Defects at the fossa ovalis (Ostium secundum):-

It is the commonest type. Its posterior border may be so deficient that the posterior atrial wall forms a boundary for the defect, separating the inferior edge of the defect from the atrioventricular valves is atrial septal tissue. By virtue of its position, the defect has its postero-inferior zone in close proximity to the right atrial orifice of the inferior vena cava. (Hurst, 1982). The anterior rim of the defect is continuous with a well-

developed Eustachian valve, a point of some practical imporatnce in repair.

Secundum ASD, may be one large defect or may consist of several small, almost cribriform openings.

The size of ASDs varies from at least one centimeter in diameter - the smallest, clinically significant opening - to virtual absence of the atrial septum, the average size is approximately 2 to 3 $cm.^2$

An adult with a defect of 2 $cm.^2$ was considered by Dexter as functioning with a common atrium. (Nadas, 1972). Defects less than 1 $cm.^2$ are usually asymptomatic. (Pomerance and Davies, 1975).

Ostium secundum defect may occur as an isolated anomaly or in association with other defects. Commonly associated defects are patent ductus arteriosus, ventricular septal defect, pulmonary stenosis, anomalous pulmonary venous drainage, transposition of the great arteries and Fallot's tetralogy. (Perloff, 1978). The Holt-Oram syndrome may be associated with secundum atrial septal defect. (Braunwald, 1980).

Various causes of left atrial enlargement such as patent ductus arteriosus, ventricular septal defect,and mitral valve disease, or right atrial enlargement as in primary pulmonary hypertension may cause tensions

or disproportions at the site of an intially valvular-competent foramen ovale, so as to yield a septal defect. (Tandon, 1974). Removal of the underlying condition may result in spontaneous closure of the defect. (Mody, 1973).

Pathological changes seen in all types of atrial septal defects with significant shunting include both cardiac chambers and pulmonary vasculature.
1- The cardiac chambers:- In uncomplicated atrial septal defect with left-to-right shunt, the two atria and the right ventricle participate in carrying the shunt. The left ventricle does not participate and remains normal. The right atrial and ventricular chambers become grossly enlarged but, their walls are not hypertrophied. The left atrium has a tendency not to become enlarged. This is explained on the basis of the assumption that as the shunted blood enters the left atrium it is not retained in this chamber, but instead flows immediately into the right atrium.

When the atrial septal defect becomes complicated by pulmonary hypertension, the right ventricular walls become hypertrophied and offer greater resistance to filling and underlie the development of a right-

to-left shunt. (Hurst, 1974).

2- Pulmonary vasculature:- In uncomplicated atrial septal defect, the major pulmonary arteries are dilated and the pulmonary trunk is considerably wider than the aorta. As the pulmonary trunk dilates,the individual cusps of the pulmonary valve are stretched. When pulmonary hypertension enters as a complication, atherosclerosis occurs. Saccular aneurysms and thrombosis of the major pulmonary arteries may occur. Dissecting aneurysm or rupture of the pulmonary trunk are among the rare complications.

Among patients with atrial septal defect and left-to-right shunt, pulmonary hypertension may develop but usually not before the third decade. In such cases, occlusive pulmonary vascular lesions are apparent. The earliest lesion appears to be characterized by cellular fibrous initial thickening in the proximal segments of the arterioles. Following the development of this process, the pulmonary arterial pressure rises; then follows the development of medial hypertrophy of muscular arteries and appearance of plexiform lesions. In the final state, the pulmonary vascular bed may be difficult to distinguish from that in ventricular septal defect with occlusive pulmonary vascular disease. (Hurst, 1982).