Balloon Dilatation Angioplasty For Coarctation Of The Aorta

Thesis Submitted In Partial Fulfillment Of The M.D. Degree
In Cardiology

By

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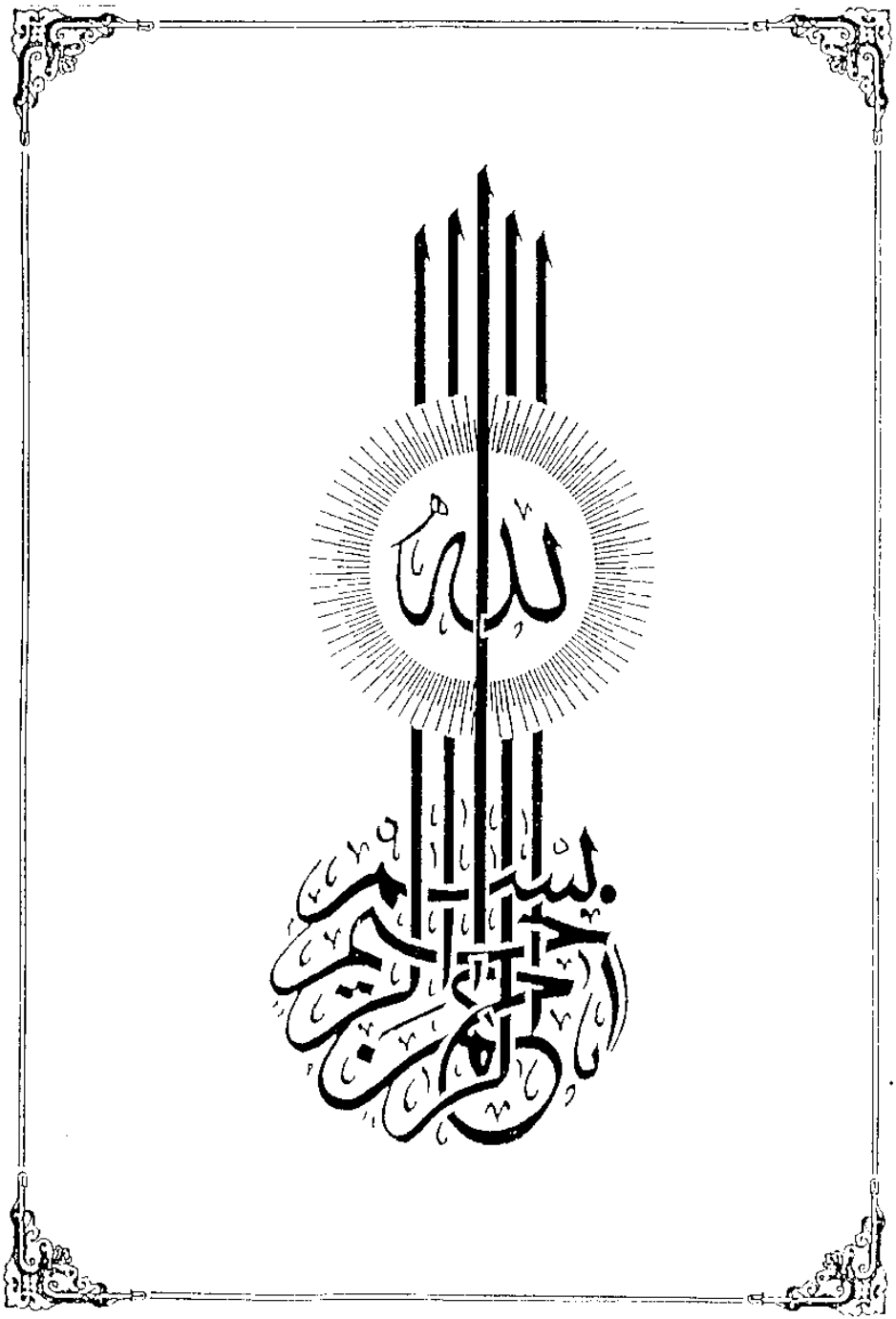

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Dedication

To My Wife,
and to My Kids Omar and Ola.

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Introduction and Aim of the Work

INTRODUCTION AND AIM OF THE WORK

Coarctation of the aorta (CoA) is a constriction of the aorta, either discrete or of a significant length, which is invariably, located at the junction of the ductus arteriosus and aortic arch just distal to left subclavian artery *(Nugent et al., 1986)*.

CoA is one of the common cardiovascular defects. It is the fifth or sixth most common of the congenital cardiovascular defects *(Keith et al., 1988)*.

CoA is widely recognized as a "treatable" form of hypertension, and patients with the defect are often identified by routine blood pressure screening. The pathognomonic clinical features include differential blood pressure between the upper and lower extremities with systolic hypertension present in the right arm or both arms. Diminished or absent femoral pulses are an additional hallmark of this defect *(Liberthson, 1989)*.

Coarctation in infants, children and young adults typically causes few or no symptoms and is diagnosed after routine physical examination. Symptoms relate to acquired complications, which include systemic hypertension, heart failure, stroke, aortic dissection, and infective endocarditis *(Liberthson, 1989)*.

The first coarctation repair was performed by *Crafoord* in 1945. In general, the results of operative management have been excellent, with low morbidity and mortality rates in uncomplicated cases.

The incidence of recoarctation in infants undergoing end-to-end anastmosis or subclavian flap angioplasty of the aorta in the first year of life is in the order of 30% ***(Cooper et al.,1989)***. If surgical repair delayed to later childhhod, there would be a risk of persistent hypertension; although the risk of recoarctation would be lessened ***(Kan et al., 1983)***. Even selected normotensive patients with good operative results have abnormalities such as residual left ventricular hypertrophy and hyperkinesia ***(Carpenter et al., 1985)***.

In 1979, ***Sos et al***, reported the feasibility of dilating CoA in post-mortem human specimen. Subsequently, excised coarctation segments obtained at surgery from infants and children were dilated in vitro. Histologic studies revealed intimal and medial tears suggesting possible mechanism for the relief of the aortic obstruction ***(Lock et al., 1982)***. Percutaneous balloon angioplasty of coarctation was first described by ***Singer et al*** (1982) and has since, been used in large numbers of patients with CoA. In virtually all, short-term success in gradient reduction and increase in CoA diameter has been reported. In majority of cases, no serious morbidity occurred. The rare mortality noted during the balloon procedure has been related either to presence of associated ductus arteriosus or to perforation of the aorta after the procedure by an angiographic catheter reinserted without guide wire in place ***(Finley et al., 1983)***.

Short-term benefits, aside from CoA gradient relief, have included relief of systemic hypertension and lack of paradoxical hypertension ***(Kan et al., 1983)***.

The long-term results, however, have been less uniformly positive. Whereas most follow-up studies report continued patency and enlargement of the aortic diameter at the coarctation site; ***(Allen et al., 1986, and Rao et al., 1987)***, a few studies ***(Lock et al., 1983, and Cooper et al., 1987)*** note an increase in the transcoarctation gradient from the immediate post-Balloon Dilation Angioplasty (BDA) decrease.

Transluminal balloon angioplasty appears to be an attractive alternative to surgical repair of CoA; most authors agree that larger series over longer periods of time are still needed for the better assessment of the real incidence of complications especially aneurysm formation ***(Samul,1989)***.

AIM OF THE WORK

The aim of this work is to study early and intermediate term results of Balloon dilatation angioplasty as a nonsurgical management of CoA.

Review of Literature

COARCTATION OF THE AORTA NATURAL HISTORY AND PROGNOSIS

"Anatomical and physiological observation concerning extra ordinary dilation of the heart which came from the fact that the aortic conduit was too narrow" introduced the description of coarctation of the aorta by the ***Prussian anatomist Johann Friederich Meckel*** in 1760.

CoA is a congenital constriction of the aorta of varying degree which is invariably located at the junction of the ductus arteriosus and aortic arch just distal to the left subclavian artery ***(Nugent et al., 1986)***. It is either discrete or of significant length.

The characteristic lesion is a deformity of the media of the aorta. The deformity involving the anterior, superior, and posterior walls, is represented by a curtain-like infolding of the wall which causes the lumen to be narrowed and eccentric ***(Nugent et al., 1990)***.

Histologic examination of the CoA ridge shows that it consists of infolding and thickening of aortic media in such a fashion that ductal tissue is incorporated into the inner part of the ridge ***(Elzenga et al., 1983)***.

The basic lesion is therefore composed of aortic media and ductal tissue, with subsequent development of a thick layer of intimal proliferation on and distal to the ridge, leading to progressive narrowing of the residual lumen. There is dilatation of the aorta immediately above, but especially just below, the coarcted segment (Fig. 1 & 2).

This morphologic feature of the anomaly was responsible for the latin term "Coarctus". which means contracted, tightened or pressed together.

The mechanism responsible for the location of typical CoA near the aortic orifice of the ductus is unresolved. The hypothesis that obstruction develops chiefly because ductal tissue in the aorta constricts during closure of the ductus is debatable because coarctation sometimes occurs with a widely patent ductus ***(Elzenga et al., 1983)*** and because coarctation has been observed to evolve in utero ***(Allan et al., 1984)***.

In symptomatic infants, the lesion lies either opposite the ductus or in a preductal location. In adolescents and adults, it is usually distal to the ligamentum arteriosum. In rare cases the lesion lies proximal to the origin of the left common carotid artery. The zone of CoA is characteristically located immediately beyond the left subclavian artery, the proximal portion of which is dilated (Fig 1-a) . Less commonly, the coarctation lies at or immediately proximal to the left subclavian artery ***(Johansson et al., 1961)***, locations that compromise the left subclavian orifice (Fig.1-b). In exceptional instances, the right subclavian artery arises distal to the coarctation and is therefore in the low-pressure zone (Fig. 1-c).

In addition to these variations, "abdominal coarctation" sometimes occurs ***(Bahabozorgui et al., 1966 & Haldane, 1983)***. Controversy continues over whether abdominal sites are congenital or acquired. The current consensus is that three principal categories exist. Some constrictions appear to represent congenital hypoplasia

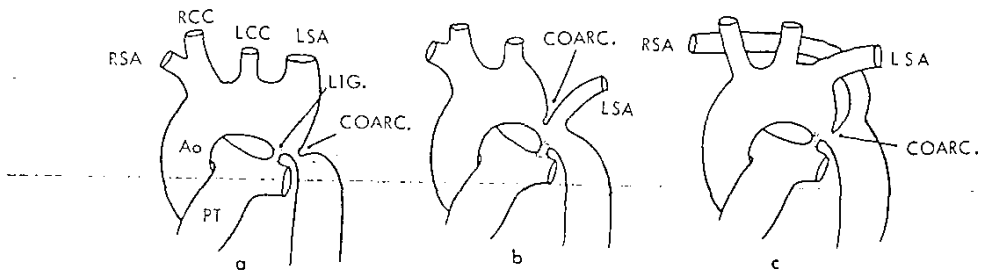

FIGURE -1. Illustrations of the typical variety of coarctation (COARC.) of the aorta and two anatomic variations. *a*, In the typical variety, the coarctation is located immediately beyond the left subclavian artery (LSA), which is enlarged. The descending aorta is dilated distal to the coarctation. (RSA = right subclavian artery; RCC and LCC = right and left common carotid arteries, respectively; LIG = ligamentum arteriosum; Ao = ascending aorta; PT = pulmonary trunk.) *b*, The site of coarctation is just proximal to the left subclavian artery. The left subclavian is not dilated. *c*, The *right* subclavian artery (RSA) arises anomalously below the coarctation.

Figure 1. Quoted From: Perloff, J.K. Clinical Recognition Of Congenital Heart Disease (3rd ed.),Philadelphia: W.B. Saunders; 1987.

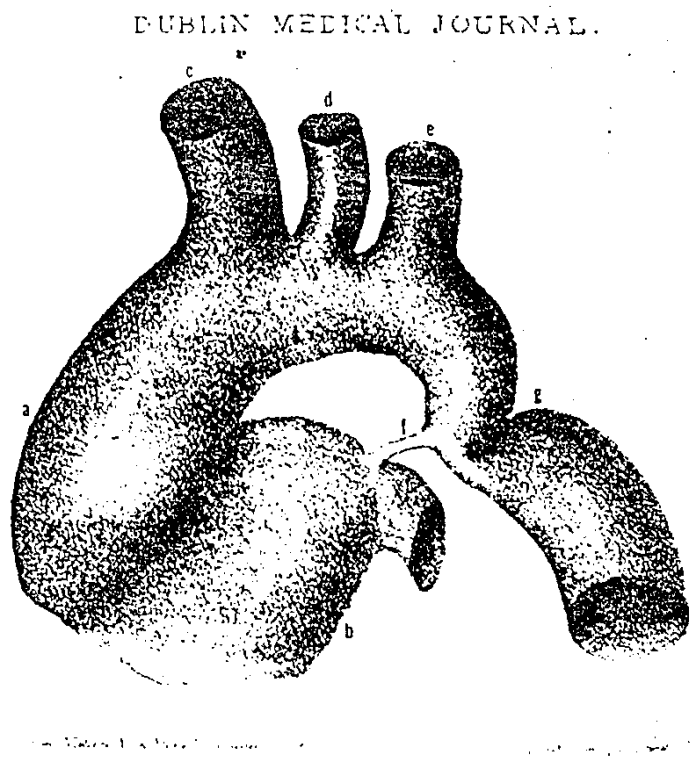

FIGURE -2. An anatomic sketch of coarctation of the aorta published in 1834. Compare with Figures 7–1*a* and 7–3. (From Nixon, R. L.: Dublin J. Med. *In* Chem. Sci. 5:386–400, 1834.) (Courtesy of Dr. Saul Jarcho.)

Figure 2. Quoted From: Perloff, J.K. Clinical Recognition Of Congenital Heart Disease (3rd ed.), Philadelphia : W.B. Saunders; 1987.

of the abdominal aorta and its visceral branches ***(Haldane, 1983)***. Intimal hyperplasia is the chief histologic finding. A second category is the acquired segmental abdominal aortic stenosis of Takayasu's disease ***(Weiner et al., 1983)***. A third category of abdominal coarctation is acquired fibromuscular dysplasia ***(Lande et al., 1976)***. Regardless of cause, abdominal coarctation is usually accompanied by serious hypertension due to renal artery stenosis ***(Haldane, 1983)***.

CoA is one of the common cardiovascular defects. It is the fifth or sixth most common of the congenital cardiovascular defects ***(Keith et al., 1978)***. Among infants, CoA ranked fourth in the New England Regional Study of Congenital heart defects, accounting for 7.5 % of all the infants with cardiac malformations under one year of age ***(Fyler et al., 1980)***. This may represent an underestimation since it has become apparent that new born infants may not display a difference in blood pressure between the upper and lower limbs in the first few days of life ***(Rudolph et al., 1972)***. The evolution of coarctation seems to be an active process in some babies, resulting in an increasing degree of obstruction and blood pressure gradient with time.

Coarctation in infancy is associated with only a slight male predominance, whereas isolated CoA in older patients is more common in males. The male to female ratio is 1.74 to 1 ***(Campbell & Polani, 1961)***, while in other series it is 3 :1 ***(Glancy et al., 1983)***.

Several congenital cardiac defects may be associated with CoA. Bicuspid aortic valve is the most frequently encountered malformation accounting for as much as 85% of cases in some series