SUMMARY

Coarctation of the aorta is a congenital narrowing of the lumen that produces an obstruction to the blood flow. This narrowing can occur anywhere from the arch to the aortic bifurcation; however, 98% occur between the left subclavian artery proximally and the junction of the aorta and ductus arteriosus distally.

The embryologic origin of coarctation of the aorta is explained by two theories. In patient with coarctation of the aorta it has been shown that the obstructing shelf is largely composed of tissue similar to that found in the ductus arteriosus. Contraction and fibrosis of this tissue at the time of ductal closure would lead to constriction of the aorta and obstruction to blood flow. It has been also proposed that coarctation of the aorta is the result of abnormal fetal blood flow patterns associated with decreased ascending aortic output.

The aortic wall including the coarcted segment histologically consists of adventitia, media and intima. It is composed of four types of tissue; namely endothelial lining, elastin fibres, collagen fibres and smooth muscles. Histopathological studies of excised coarcted aortic sements showed abnormalities in the elastic tissue described as cystic medial necrosis and defined as depletion and disarray of elastic tissue. Variable degrees of elastic tissue abnormalities were observed. Medial abnormalities were identical proximal to the coarctation (high pressure and low velosity zone) and distal (low pressure and high velosity zone) inplying that the abnormalities were not haemodynamically determined.

Coarctation of the aorta accounts for 8% of congenital heart disease. The association of a bicuspid aortic valve with coarctation of the aorta is well recognized as it ranges from 25 to 85%. Neonatal aortic coarctation is frequently associated with hypoplasia of the transverse arch. It may be associated with patent ductus arteriosus, ventricular septal defect and atrial septal defect.

Nearly one half of patients with coarctation of the aorta develop symptoms during the first months of life. Without collateral decompression these infants present with tachypnea, poor feeding, hepatomegaly, cardiomegaly, cyanosis and congestive heart failure. Upper extremity hypertension may exist with lower extremity hypotension. Differential cyanosis may exist between the upper and lower extremities. Generalized systemic hypotension, oliguria, renal failure and severe metabolic acidosis with ultimate death may ensure without intervention.

The chest radiograph shows significant cardiomegaly and the electrocardiogram shows sinus tachycardia and biventricular hypertrophy. Echocardiography will confirm the diagnosis and associated intracardiac lesions should be suspected.

MRI also compares well with angoigraphy for evaluating both the site and the severity of the coarctation as well as the occurrence of collateral vesells and aortic dimensions.

Balloon angioplasty is generally considered safer in patients with postsurgical recoarctation (because of the supporting effect of surrounding scar tissue) rather than native coartation.

Stenting has been gaining acceptance in the treatment of isolated coarctation of the aorta. Its theoretical advantage over simple balloon dilatation is a potentially lower incidence of recoarctation.

Three types of surgical repair remain in common use: resection of the stenosed segment of aorta with end to end anastomosis, use of subclavian flap and patch aortoplasty (using synthetic material for patch).

Mortality rates, residual or recurrent coarctation rates, and complication rates do not provide definitive information on which to base recommendations for exclusive use of any of the various methods of repair. Each technique has advantages and disadvantages, some theoretical and others practical. Familiarity with the various techniques and awareness of specific advantages and disadvantages of each, coupled with a clear understanding of the individual patient's morphology and physiologic status, allows the surgeon to make the best possible decision regarding the timing and technique of surgery. The most important principle is that the surgeon should not take an intellectually rigid approach.

At the present time it is believed that some form of resection with anastomosis is the procedure of choice under the following conditions: the patients is physiologically stable when taken to the operating room, and there is the potential for complete resection of ductal tissue and anastomosis proximal to all significant tubular hypoplasia without

producing tension on the suture line. The availability of prostaglandin E_1, the development of absorbable suture material, the attractive concept of completely removing the abnormal tissue from the aortic lumen, and the experience gained from other procedures (e.g., arterial switch operation) demonstrating the circumferential tension-free aortic suture lines invariably grow normally when normal tissue is involved, all support this approach. If these criteria cannot be met with confidence, another repair technique should be chosen, based on specific morphologic and physiologic consideration.

Recommendation regarding the timing of coarctation repair cannot be made with certainty. At the present time it is believed that elective coarctation repair is best performed at 3 to 6 months of age and repair in symptomatic patients should be undertaken at the time of diagnosis.