Kinking of Aortic Arch with Aneurysmal Dilatation

Report of a Case

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KINKING OF THE AORTA IS A RARE anomaly in which the aortic arch is kinked in the shape of letter S. This congenital anomaly has been described and established as a clinical entity in the last few years. Although usually presenting a characteristic radiologic appearance, it is not as yet widely appreciated either by radiologists or by clinicians.

Rössler and White in 1931 first described the uncommon finding of an elongated aorta. The clinical description of this entity was first reported by Soudas et al. in 1951. The term "kinking of aortic arch" introduced by DiGuglielmo and Guttadauro aptly described tortuosity and curvature of the elongated aortic arch. To date, about 40 such cases have been described in world literature. One such patient seen at the cardiology department of the B.Y.L. Nair Hospital, Bombay, is the subject of this report.

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CASE REPORT

M.J., a 28-year-old housewife, was seen at the B.Y.L. Nair Hospital for left-sided non-anginal chest pains and occasional bouts of cough and giddiness since childhood. Past and family histories were non-contributory.

Physical examination revealed a healthy woman in no acute distress. The temperature was 98.6°F., the respirations 16 per minute and the pulse was 80 per minute and regular. The left sided brachial and radial pulses were weaker as compared to right. The other arterial pulses were equal bilaterally. The blood pressure was 110/70 mm Hg in the right arm as compared to 90/70 mm Hg in the left arm. The blood pressure in the legs was 140/96 mm Hg. The optic fundi were normal. The jugular venous pressure was not elevated.

Examination of the heart showed an apical impulse in the fifth left interspace inside the midclavicular line. No abnormal pulsation was seen or felt. Heart was not enlarged on percussion. On auscultation the heart sounds were normal. The second sound over the pulmonary area was normally split. A grade 2/6 ejection systolic murmur was heard over the left second and third intercostal spaces. No diastolic murmur was heard. Results of the balance of the physical examination were unremarkable.

Laboratory Investigations

The routine blood counts and urinalysis were unremarkable. Serologic test for syphilis (blood V.D.R.L. test) was negative. The electrocardiogram was interpreted as normal. The X-ray films of the chest (Fig. 1) revealed a soft superior mediastinal shadow and low placed aortic knuckle on the left side. The heart size was normal. The lungs were clear. No calcification was observed. On fluoroscopy, the left superior mediastinal shadow showed systolic expansile pulsations and was thought to be the kinked aortic arch. Barium swallow in the posteroanterior view of the chest demonstrated displacement of the esophagus to the right with a generalized concave contour to the left, and in the left anterior oblique view, there was anterior displacement of the esophagus (Fig. 2). An aortogram (Fig. 3) performed through the right brachial artery showed a normal sized ascending aorta, and kinking of the aortic arch with aneurysmal dilatation.
of the kinked segment. This kinked segment had a double S shaped curve. The aorta was slightly narrowed just proximal and distal to the kinked portion. The descending aorta—the post-kinked

**Figure 2**: Barium swallow showing displacement of the esophagus. (A) Right anterior oblique view showing posterior displacement of the esophagus. (B) Posteroanterior view showing rightward displacement of the esophagus. Note the generalized concave contour to the left. (C) Left anterior oblique view showing anterior displacement of the esophagus.

**Figure 3A**: Aortogram showing kinked aortic arch and aneurysmal dilatation. Note the slight narrowing of the aorta proximal and distal to the kinked segment. The post-kink segment is slightly dilated.
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segment — was minimally dilated. The aortic arch was left-sided and the descending aorta lay on the left of the midline. The brachiocephalic vessels appeared normal.

**DISCUSSION**

Blake and Manion have stated that congenital elongation or “looping” of the aorta may be caused by multiple alternating involutions affecting both the aortic arches. Barry described the complex changes occurring in the embryonic aortic arch development and postulated that they may explain such aortic arch anomalies. During organogenesis some arterial segments elongate while others contract. Such changes occur in the seventh dorsal segmental arteries where they undergo a cephalic shift and ultimately become the origin of subclavian arteries. Compression of the third to seventh dorsal aortic root segments and of the fourth arch segments occur with the descent of the heart into the thorax. During this development, the origin of the subclavian arteries remain relatively far cephalad. Failure in compression of the above mentioned segments during development results in an abnormally long and high placed aortic arch which “twists” or “kinks” at the point of insertion of the ligamentum arteriosum. The kink lies to the front and somewhat to the right at the level of ligamentum arteriosum, where aortic coarctation most frequently occurs. The term “pseudocoarctation” was thus used for this anomaly. This may, however, be distinguished from true coarctation of the aorta by the absence of hemodynamically significant stenosis and collateral circulation. The pulsating oval opacity situated higher up on the left superior mediastinal border represents the proximal portion of the kinked aortic arch. The apparent aortic knuckle is the end on view of the descending aorta distal to the kink. Pattinson and Grainger state that the apex of this abnormal kink is closely applied to the upper and posterior aspect of the left pulmonary artery and left main bronchus, which may be displaced forward, downward and to the right. The esophagus, also being closely related to the kinked segment, is displaced forward and to the right. Such was the case in our patient. Clinically, the aortic closure sound may be loud because of increased aortic curvature displacing the ascending aorta closer to the anterior chest wall. The ejection type systolic murmur present in our case is probably due to turbulence of the blood flow at the sudden bands on the aortic arch.

Diagnosis of kinked aortic arch can be made by chest x-ray films. The posteroanterior chest film shows double opacity, the upper one less opaque than the normal aortic arch, lying along the left superior mediastinal border. The lower opacity is the end on view of the post-kink segment. The barium swallow reveals double concave indentations of the esophagus, the upper one due to the kinked segment and the lower one secondary to the post-kink portion. Sometimes a long continuous concavity combining the two may be seen.

Aortogram performed preferably through the right brachial artery clinches the diagnosis. Slight narrowing at the site of kinking and dilatation distal to the kink are not uncommon. Aortogram in our patient demonstrated such features and also showed aneurysmal dilatation of the kinked segment. Pressure effect of this dilated kinked segment on the left subclavian artery origin would account for the discrepancy in the

**Figure 3B:** Diagrammatic representation of the aortogram.
arterial pulses and blood pressure of the upper extremities in our case. Saric et al. have suggested that these patients may develop aneurysmal dilatation because of the pounding of the blood stream against the bands of the tortuous aortic arch. The onset of atherosclerosis may increase the impending danger of aneurysm formation.

It is true that in some cases of kinked aortic arch, organic heart diseases have been present. Patent ductus arteriosus, ventricular septal defects, dilatation of aortic sinuses of Valsalva, aortic valvular stenosis and endocardial fibroelastosis have been described as associated lesions. Our patient did not have any evidence of associated lesion.

Kinking of the aortic arch and of descending thoracic aorta simulate intrathoracic tumors and so it is imperative to make the proper diagnosis to avoid fruitless and potentially hazardous thoracotomy. Surgery has so far not been undertaken for this anomaly and we did not feel it indicated in our case.

INFLUENCE OF STROPHANTHIN ON DURATION OF CARDIAC CYCLE PHASES

Strophantin in a dose of 0.25 mg. was given intravenously to 30 patients suffering from cardiac diseases with diverse degrees of circulatory insufficiency. Before the administration and one hour thereafter, the authors recorded electrokymograms of the left and right regions of the heart, on the basis of which, subject to separate analysis were the systolic and diastolic phases of the left and right ventricles. The myocardial action of strophanthtin on the left and right cardiac regions is identical and does not depend on changes of the rate of cardiac contractions. Strophantin intensifies the systolic action, not only of the ventricular muscles, but also of the atrial myocardium. Its systolic effect is manifested by a shortening of the contraction period and alteration of the ejection period. In mitral stenosis, the period of ejection becomes prolonged after the administration of strophanthtin. In other valvular affections of the heart, it becomes shortened. The diastolic action of strophanthtin is manifested by a shortening and deepening of the period of diastole.


CHRONIC ANEURYSMS OF THE HEART

Under study were 115 patients with chronic aneurysms of the heart. This group comprised 9.5 per cent of patients who have sustained myocardial infarction. Clinically chronic cardiac aneurysms were diagnosed in half the cases. Among the causes which hampered diagnosis, the author noted the following: faulty anamnesis in patients with marked cardiac failure, incomplete clinical examination of patients, indistinct symptoms in the localization of small aneurysms on the posterior wall. The most characteristic signs of chronic aneurysm of the heart are: auricular pulsation, systolic and diastolic murmurs, cardiac insufficiency. Among the causes of death, the author notes cardiac insufficiency, thromboembolization, stroke peritonitis and uremia.