Long Survival in Dissecting Aneurysm of the Ascending Aorta*

By

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Dissecting aneurysm of the aorta, more recently known as aortic dissection is a highly lethal condition.[1] With the availability of improved diagnostic modalities, medical and surgical therapy since 1960, the survical has improved significantly.[2-3-4]

This has lead to thorough search for this disease by the clinician resulting in increased recognition of this once common autopsy diagnosis[5]. Approximately two thousand new cases are diagnosed in USA each year[6]. In Pakistan this disease is extremely rarely diagnosed if ever confirmed. We report here two documented cases of dissecting aneurysm of the ascending aorta diagnosed in NICVD in the past one year with long survival.

Aortic dissection is an extremely fatal condition resulting in the loss of life of the affected person with great rapidity. It has been estimated that the Atrition rate for Ascending Aortic Dissections (A.A.D.) is 1% per hour for the first 48 hours and the majority are dead in the first 3 weeks if appropriate therapy is not institututed. The mortality for the descending aortic dissection is relatively less.

Table No. 1 gives details of the various types of aortic dissections with their distinctive clinical and haemodynamic features.

DEBAKEY TYPE	AORTIC INVOLEMENT	DAILY TYPE	AORTIC INSUFFICIENCY	TAMPONADE — HEMOTHORAX	CORONORY OBSTRUCTION & DISECTION	CEREBRAL VESSEL OBSTRUCTION	ABDOMINAL VESSEL INVOLEMENT	THERAPY
I	ASCENDING AORTA · ARCH · DESCENDING AORTA	A	+ +	+ +	±	+ +	+	A1 A2
II	ASCENDING AORTA	A	+ +	+ +	±	+ +	−	
III	DESCEND. ING AORTA DISTAL TO LT. SUBCLAVIAN ARTERY	B	NIL	NIL	NIL	NIL	+	B1 B2

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The classification of DeBakey et al presented in 1965, groups cases of A.A.D. into 3 categories. Type I where the initimal tear is situated just above the Aortic valve where the dissection starts and spreads across the arch into the abdominal aorta.

Type II the dissection is limited to the ascending aorta.

Type III the dissection begins just distal to the left subclavian artery and involves the thoracic and abdominal aorta for variable distance. Because of the inability to localize the initimal tear in about 10% cases, presence of tear in the arch of the aorta, retrograde spread of type III to the arch of the aorta and the virtual similarity of Type I and Type II as regards treatment, complications and prognosis, Dailey et al simplified the classification by dividing all the cases into Type A and B. Type A include all cases of A.A.D. involving the ascending aorta. Type B include all those cases not involving the ascending aorta.

More recently Braunwald has used the term proximal for all cases involving the segment of aorta proximal to the left subclavian artery and distal for all other cases.

CASE REPORTS

Case No. 1:

A.K. 40 years old Male refered to N.I.C.V.D. for evaluation of aneurysm of the Ascending Aorta and recurrent chest pains of 2½ years duration. The patient who works as a Peon in Government Office, was doing well until 2½ years ago when he developed severe "tearing" mid sternal chest pain radiating to the inter scapular region associated with profuse sweating, and vomiting, he was taken to the hospital where his B.P. was found to be 60-70 mm Hg. He was kept in the hospital for 10 weeks and then discharged. Four months following this the patient had another similar episode and was hospitalised. It was found that he has aortic insufficiency and also ascending aortic aneurysm. He was noticed to be in L.V.F. and improved with Digoxin and diuretics. He was advised surgery but refused, since then patient complains of dyspnea and retrosternal chest pain on exertion and 15 days prior to admission patient had another resting pain with sweating. He was sent to NIVCD for evaluation. There is no history of hypertension, Rh. fever and syphilis. On physical examination patient was well developed well nourished male in no acute distress. Height is 5' 3" weight 147 lbs. no stigmata of marfan syndrome apparent. B.P. (R) arm 120/65 left arm 115/70 JVP flat thyroid not palpable P.M.I. 6th ICS AAL: S1 S2 II/VI early Diastolic murmur L.S.B. lungs clear to A&P, liver, spleen kidney not palpable pedal edema nill peripheral pulses normal C.N.S. unremarkable, ECG normal sinus rhythm 85 min. regular P.R. interval .16 sec. QRS .06 sec. axis+30 ST depression and T wave inversion V2 to V6 consistent with subendocardial ischaemia and or infarction. Chest X-ray P.A. cardiac silhouette enlarged, consistent with ascending aortic aneurysm or unfolding; V.D.R.L. negative. Aortogram was done on 23-2-82 from the right femoral with 8F pigtail catheter; left lateral; left anterior oblique and A.P. views. 76% Renografin 50 cc dye injected 25 cc/sec. and recorded on 35 mm cinefilm on Siemen Cardoscop. Review of the film revealed a sizeable intimal tear in the ascending aorta approximately 3 cms above the aortic ring; the true lumen was compressed and the false lumen was huge resulting in dilatation, distortion and dislocation of Aortic ring and severe A.I.; the false lumen

extended to the right Brachiocephalic vessel, but not obstructing it, no significant clot was seen in the false lumen Diagnosis of type II Dissection was confirmed.

Patient underwent successful surgery with aortic valve replacement and ascending aortic graft at NICVD.

Case No. 2:

S.A. 36 years old male known to have had border line high blood pressure (140/90), since 1968 was admitted to NICVD on 12th November, 1980 with the diagnosis of gross pulmonary oedema of one week duration. Patient was doing well till one week prior to admission when climbing stairs in Cairo noted severe crushing retrosternal pain followed rapidly with palpitation and shortness of breath. Tne pain lasted one hour and was eased with some injection. However soon after he became very restless, started coughing accompanied with frothy blood stained sputum. The patient was advised admission but he became extremely confused and wanted to be brought to his home town in Pakistan. He was flown to Karachi, where on arrival he was seen by a family physician and started on antituberculous medication. However his condition became worse and he was sent to NICVD where on examination he was found to be acute in respiratory distress, JVP was raised B.P. was 160/60; thrill was palpable along LSB there was grade 5/6 ejection systolic murmur alongwith grade 5/6 early diastolic murmur. Heart sounds were buried in the murmur. Bilateral rales were audible; Liver + enlaged; oedema feet nil; peripheral pulses were bounding. His height was 5 feet 8 inches, and weight 122 pounds, finger joints were lax but no overt stigmata of Marfans.

He was given heavy doses of I.V. Lasix and digitalised. Patient settled and had an Echo examination which was suggestive of grossly dilated aortic root as well as acute AI with early M.V. closure. No vegetations were seen. Patient underwent aortography and was found to have grossly dilated aneurysmal ascending aorta with gross AI; an intimal tear could be clearly visualised four centimeters above the Aortic valve. The dissection was confined to the ascending aorta. Patient was offered surgery which he refused.

He was started on antihypertensive treatment to which he responded well. However, after one year he was operated on at Toronto on November 27, 1981. The diagnosis of Aortic dissection ascending aorta (Type II) was confirmed. He underwent aortic valve replacement and ascending aortic graft with reimplantation of both coronary arteries.

Discussion:

Najafi uses the term Anterior for the proximal and posterior for distal dissections. As shown in figure I(A) the intimal tear is very well seen

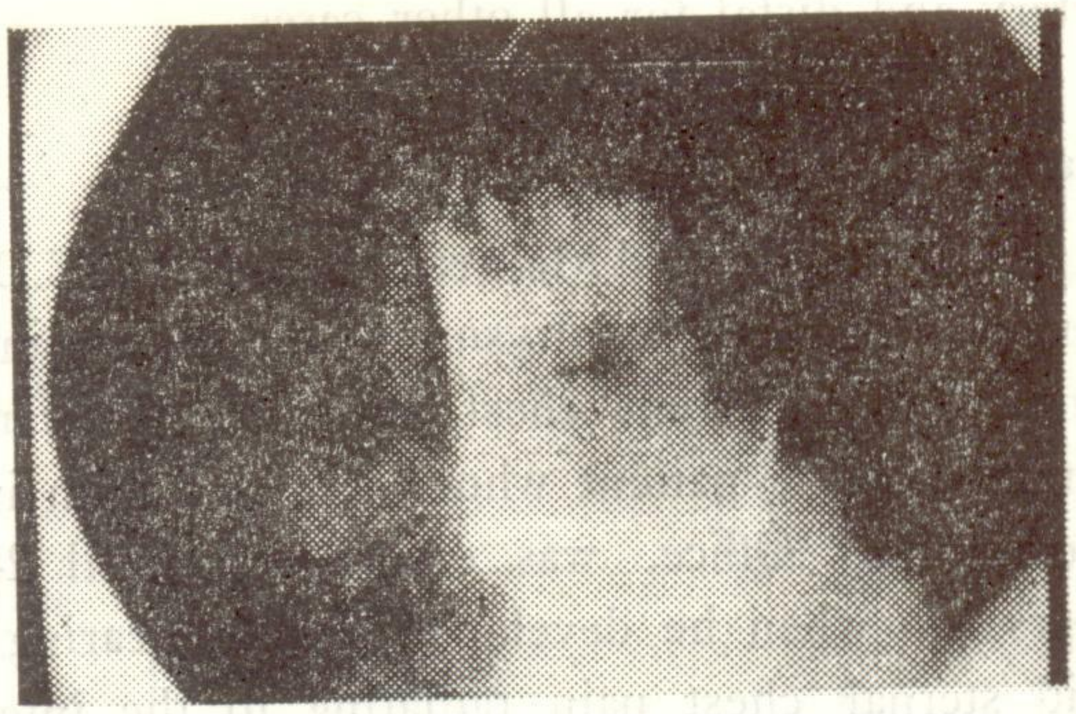

Fig. I (A)

just distal to the aortic valve and dye is seen entering into the huge, dilated false lumen.

Fig. I (B)

In Figure I(B) the true lumen is compressed greatly by the false lumen. The dissection is not encircling the lumen completely and as shown in Figure I(C) both the coronary arteries are spared. Gross A.I. is present and marked volume overload of L.V. resulting in L.V.F. controlled with digoxin and diuretic, in this patient. Thus both our cases belonged to Type II of DeBakey classification. Inspite of huge, patent, false lumina in both our cases such long survival is exceptional. None of our cases had typical occular or somatic stigmata of marfan's syndrome and mild systemic hypertension was present in one case only. It is interesting to note that aortic insufficiency secondary to marfan's syndrome is frequently seen in this country. Yet Dissection has not been reported in these cases. The other major category in which Dissection occurs is systemic hypertension. It has been reported that systemic hypertension is as common in Pakistan as in the West.[7] Coarctation of the aorta and aortic valve disease are also commonly encountered in this country however dissection has not been reported in this group either. Shanwar in 1975[8] wrote about the aortic dissection, "This disease has long been misdiagnosed, misunderstood and frequently mis-interpreted". In both our cases the diagnosis came as surprise at the time of aortography for evaluation of A.I. in one case, and A.I. and Aortic anneurysm in the other case. One of our case was treated for Tuberculosis. However, L.V.F. due to A.I. was diagnosed when a Cardiologist was consulted. Both cases survived more than few months before the diagnosis was established. Keeping in view the life history of Acute Dissection, the index of clinical suspicion, nonavailability of aortogram in acute emergency situation are some of the factors, mainly responsible for the non-diagnosis of this disease in our country.

The aortic media mainly consists of collagen, elastin and smooth muscle cells, giving it tensile strength against rupture, elasticity and shock absorbing properties, so essential for aorta, to function as a conduit for pulsatile blood flow. The inner one 3rd of this medial layer is avascular and highly depends upon the distant vasavasorum and the blood in the aortic lumen. Degeneration of the aortic media; occasional bleeding from the vasa vasroum and tear in

Fig. I (C)

the intima allowing blood to enter in the degenerated media may set the stage for the Dissection to start while the shearing forces of blood passing past the intima, rapidly expansile blood flow entering the aorta from a hyperkinetic L.V. and elevated mean aortic pressure may all perpetuate the process of Aortic Dissection. While the exact mechanism of aortic Dissection is unknown; (A) Degeneration of Aortic media (B) Intimal tear (C) Occlusion and Bleeding from vasa vasorum (D) Hydraulic forces, are some of the most important factors for production and perpetuation of this disease. Thus Marfan's syndrome with its associated cystic medial necrosis, systemic hypertension, coarctation of the aorta and pregnancy are all known to be associated with Acute Aortic Dissection. These factors and conditions are summarized in Table No. III. Acute Aortic dissection produces typical set of signs and symptoms summarized in Table No. IV.

Table IV. Symptoms and Signs of Acute Dissection (Modified from Anagnostopoulas)

Symptoms	*Physical findings*
(A) C.N.S.	
1. Pain in: Chest, Submental, Substernal, Facial, Epigastric, Interscapular, Neck, Midback, Sacral, Extremities	1. Hypertension in 90%
2. Syncope	2. "Shocky" cold and clammy.
3. Coma, confusion, headache	3. L.V.F.
4. Blindness	4. Unequal pulses
5. Paralysis,	5. Absent pulses
6. Hoarsesness	6. Dry, warm, pulseless extremities
(B) Cardio respiratory	7. Duplication of pulses
1. Dyspnea, orthopnea	8. Reappearing pulses
2. Hemoptysis	9. Tamponade
(C) G.I. system	10. Unilateral jugular venous distension
1. Nausea and vomiting	11. Cardiomegaly
2. Melena, hematemesis, tenesmus	12. Left hemothorax
(D) Renal	13. Pericardial friction rub
1. Oliguria, hematuria, anuria	14. Aortic insufficiency
	15. Hemiplegia, paraplegia
	16. Facial paralysis
	17. Horner's syndrome
	18. Pulsating sternoclavicular joint
	19. Acute abdomen

As will be seen Acute myocardial infarction is the main differential diagnosis. Other clinical settings where dissection should be suspected are Cerebrovascular Accident, Pulmonary Embolism and less frequently acute abdomen. If in the above set up ECG is normal, pulses are unequal, unilateral Jugular venous distension is present, sternoclavicular joint is pulsatile or Aortic Insufficiency is found, Acute Aortic Dissection must be looked for. It has been estimated that 90% cases of Aortic dissection have moderate or severe hypertension at the time of their presentation. Table No. V shows the therapeutic ABC classification proposed by Anagnostopoulos. Indications for medical and surgical therapy are summarized in Table VI and VII. It is agreed by most of the workers that unless cardiac tamponade is present, hypotensive therapy should be started as soon as strong suspicion is present for the diagnosis of Acute Aortic Dissection. Propranolol is the drug of choice and is used with trimetharphan, methyldopa etc.

Table V. The ABC Classification by Anagnostopoulos et al 1975

Class	*Treatment*
(A) Ascending Aorta (A.A.)	
A-1 With Complications	—Medical—Earliest Surgery.
A-2 Without Complications	—Medical—Planned Surgery.
(B) Does not Involve A.A.	
B-1 With Complications	—Medical—Early Surgery.
B-2 Without Complications	—Medical—Elective Surgery?
(C) Inoperable	—Medical?

Table VI. Indications of Medical Therapy

(1) Stable Distal, (Type III, B2) Descending Aortic Dissection.
(2) Clotted False Lumen.
(3) Intimal Tear cannot be Localized on Angiography, and A.I. is not present.
(4) All Hypertensive or Normotensive cases Till Properly Investigated and Final Decission for Definitive Therapy is made.

Table VII. Indications for Definitive Surgical Therapy in Aortic Dissection.

1. Proximal dissection.
2. Distal dissection complicated by:
   (a) Progression with vital organ compromise.
   (b) Rupture or impending rupture (saccular aneurysm formation)
   (c) Aortic regurgitation.
   (d) Inability to control pain or blood pressure medically.

As seen from these tables controversy still exists on the exact time of surgery and type, requiring surgery[9]. Total Medical management is possible only in those cases with distal dissection and no complications. Drugs which reduce blood pressure and left ventricular contractility are the main stay of treatment these days. The results of surgery depend upon acuteness of the process and replacement of the aortic arch versus replacement of the ascending aorta. In the series of Cachera et al[9] there was 0% hospital mortality for stable patients while 24% for acutely ill patients. The presence of shock, tamponade, C.V.A. or renal failure did not correlate with hospital mortality[10].

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