
Prolonged survival in aortic haematoma
Diagnosis and rational therapy

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Two cases of dissecting haematoma of the aorta have enjoyed unusually long survival without treatment. In both, there is angiographic evidence of a double barrel aorta, and both are asymptomatic, the one 18 and the other 7 years after the acute episode. Since the natural history of chronic aortic dissection (whether spontaneous or due to hypotensive therapy) is not well known, the circumstances that led to these fortunate results are identified. The triad of hypertension, aortic incompetence, and diminished pulsation in the right arm and right carotid arteries should lead to a strong clinical suspicion of chronic aortic haematoma. It is suggested that if angiography demonstrates flow in the false lumen, a conservative policy may be justified. Hypertension should be controlled. An operation should be considered for recurrence of acute dissection, for progressive enlargement of the aneurysm, for occlusion of major aortic branches, and for cardiac failure resulting from aortic incompetence. On the other hand, a haematoma that is arrested but not decompressed may require operative repair unless serial radiography suggests progressive resorption and fibrosis of the aortic haematoma.

Dissecting haematoma of the aorta is often a most dramatic complication of arterial disease. Untreated, the mortality may be as high as 95 per cent within 6 months. Rarely the process may become arrested or even completely healed. Should the haematoma communicate with the aortic lumen at two distinct points, spontaneous internal decompression may take place, resulting in the formation of a double barrel aorta, which is not necessarily incompatible with a normal life. In a masterful essay on aortic dissection, Burchell (1955) noted that though advances had been made in diagnosis, progress in treatment had been modest. Soon thereafter, bold surgical attack showed that in some hands, a respectable survival rate could be obtained in selected cases (DeBakey, Cooley, and Creech, 1955; DeBakey et al., 1965) and aortic dissection became a 'surgical disease'. Others, notably Wheat et al. (1965) and Wheat and Palmer (1968), have pioneered a hypotensive 'medical' regimen, reserving surgery for certain complications. The results are very encouraging.

The question then arises as to whether patients who have successfully passed the acute stage without an operation should have elective surgery to restore anatomical normality, for the natural history of the healed or partially healed aortic dissection is not well known. This paper presents two cases of exceptionally long survival after spontaneous recovery from a dissecting haematoma of the aorta, explores the factors responsible for the favourable outcome, and considers possible rational approaches to therapy.

Case reports

Case 1 In August 1953, a 50-year-old grocer suffered a short episode of severe retrosternal pain, radiating to the neck and jaw, of dramatically sudden onset. Three days later, a sudden desire to defaecate heralded a tearing pain in the front of the chest, radiating to the left scapula. Within 15 minutes the pain had spread down the left side of the back into the left hip, and the left leg became numb. Sweating was profuse.

On admission to the Veterans Administration Hospital, Wood (Milwaukee), Wisconsin, he was ashen grey, short of breath, and covered with sweat. The pulse was 120 beats a minute; blood pressure was 130/80 mmHg in the right arm and 150/80 mmHg in the left. The left femoral pulse

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was barely palpable, the right was weak, and all
distal pulses in both legs were absent. The heart
was not enlarged. A loud harsh ejection systolic
murmur, maximal at the second right interspace,
was transmitted to the apex and neck. Diastolic
murmurs were absent. Crepitant rales were heard
at both lung bases. Electrocardiogram and chest
radiograph were normal. Stigmata of Marfan's
syndrome were absent. A firm clinical diagnosis
of dissecting aortic aneurysm was made. The next
day pain in the lower back transiently increased,
migrated to the left hip, and ceased completely
after 3 days. A bruit over the abdominal aorta
appeared. On the fourth day the urine for the first
time began to show protein and red and white
cells, which persisted for 4 days.

Pulses in the right leg returned on the second
hospital day, but those in the left remained
diminished. The arm blood pressures became
equal after a week. A low grade fever, with slight
polymorphonuclear leukocytosis and high ery-
throcyte sedimentation rate persisted for 3 months.
A soft immediate diastolic murmur of aortic in-
competence along the right sternal border was
first heard during the seventh week, and a definite
widening of the aortic shadow could be seen in
the chest radiograph. After a slow and uneventful
recovery he returned to full-time work, carrying
out moderately heavy physical duties for up to 14
hours daily without difficulty.

Thirteen years later, in 1966, he sought treat-
ment for an inguinal hernia, and was referred
for an opinion because of his abnormal chest
radiograph (Fig. 1). He had no symptoms refer-
able to the heart. All peripheral pulses were
present, equal, and rather brisk. Both arm blood
pressures were 150/50 mmHg. The heart was
moderately enlarged, the apex beat showing a pro-
nounced heave. The systolic murmur was not
changed but the aortic diastolic murmur was
longer and louder. The second sound was loud.
An abdominal aortic bruit could still be heard.
The electrocardiogram showed left ventricular
hypertrophy and strain.

He remains symptom free and working full
time in 1971, 18 years after the initial episode.

Cardiac catheterization (1966)
Right-sided pressures were normal, while that
in the ascending aorta was 148/37 mmHg. A
retrograde aortogram was obtained from the
right femoral artery. Two of the serial films are
shown. In Fig. 2 (2 seconds after injection), the
natural lumen of the ascending aorta is seen in the
midst of a large shadow, and the right coronary
and brachiocephalic vessels are outlined. In Fig. 3
(at 2.5 seconds) contrast has entered the false
lumen through a rent above the right coronary
artery. The thin radiolucent septum is well seen.
Refux of contrast into the left ventricle indicates
aortic incompetence.

A second aortogram was obtained with the
catheter tip in the descending aorta. The first
film (Fig. 4) shows the true lumen narrowed be-
low the eleventh thoracic vertebra. A thin oblique
line (arrowed) represents calcium in the outer
wall of the false lumen, confirmed by tomography
(Fig. 5). All the branches of the abdominal aorta
could be identified in later films, filled from the
true aortic lumen.

Comment
The initial phase of this man's illness presents a
classical description of acute dissecting haema-
changes in serum enzymes or in the electrocardiogram. A diagnosis of myocardial infarction with coexisting rheumatic aortic valve disease was made. He made an uneventful recovery, but complained of fatigue for some months. He retired from work on medical advice, but has been able to perform heavy household chores, including mowing the lawn and removing the snow without difficulty. He has no symptoms.

In March 1970, he was referred for evaluation of the murmurs. At this time he appeared well, with no stigmata of Marfan's syndrome. In the left arm, the blood pressure was 170/70 mmHg and a waterhammer sign could be elicited. In the right arm, the blood pressure was 130/70 mmHg, with no waterhammer sign, and the pulse was clearly delayed. The pulse in the right common carotid artery was visibly and palpably less than that in the left. The external carotid pulses were also less on the right side. The two common carotid pulses are displayed in Fig. 6. The right femoral pulse was more forceful than the left, and a Durozied sign could be elicited only in the right. The left popliteal pulse was almost impalpable, but the pulses in the feet were equal. Thus, the pulses in the left arm and carotid, and those in the right leg, are suggestive of aortic incompetence, whereas those in the right arm and left leg are diminished and delayed.

Case 2 In June 1964, a 55-year-old labourer suffered a sudden, severe, tearing pain in the neck and jaw, which soon moved to the right anterior chest. Sweating and nausea were absent. After admission to another hospital the pain was relieved by morphine, and did not recur. The blood pressure in the arms was 170/110 mmHg. Leg pulses were not described. A rough ejection systolic murmur was noted on admission. An early diastolic murmur was heard in the second week. No other physical abnormality was found. The chest radiograph and the electrocardiogram were within normal limits on admission. Myocardial infarction was not confirmed by serial
Moderate cardiomegaly was present, the apex beat having a pronounced heave. Heart sounds were normal. A systolic thrill was felt and an injection murmur heard over the right sternal border, radiating to both carotids, but louder in the left. A long aortic diastolic murmur was audible all over the praecordium. There was an abdominal aortic bruit. The electrocardiogram showed left ventricular hypertrophy and strain. The chest radiograph (Fig. 7) shows cardiomegaly and prominence of the ascending aorta.

**Cardiac catheterization (1970)**
The pressure in the ascending aorta was 175/65 mmHg; the left ventricular end-diastolic pressure was 20 mmHg. Coronary cinearteriograms were normal. Retrograde aortograms were obtained. Fig. 8 shows the true and false lumens separated by a radiolucent septum. Contrast is passing into the aneurysm through a small rent in the lower part of the septum. The left ventricle is opacified because of the aortic reflux. Note that the innominate artery is narrowed as it passes through the false passage, to balloon out later. This is analogous to post-stenotic dilatation. Fig. 9 shows the obvious but regular narrowing of the true aortic lumen, from which all the major aortic branches take origin.

**Comment**
It is virtually certain that the illness of 1964 was the acute phase of aortic dissection. The clinical diagnosis at that time of acute myocardial infarction together with rheumatic aortic valve disease, a common error in this condition, cannot be sustained. The description of the pain, the absence of electrocardiographic and enzyme changes, and normal coronary arteriograms make the diagnosis untenable. This man, like Case 1, suffered an aortic haematoma which spontaneously decompressed by gaining entrance to the aortic lumen at two points, and made an uneventful recovery.

The innominate artery is involved in the dissection process, which surrounds the vessel like a cuff. The lumina of the brachiocephalic vessels are otherwise normal. The unusual physical signs of blunting of the right carotid pulse and diminution of the right arm pulses must be associated with this cuff. It is probable that the peak of pressure in the false lumen, occurring later than in the true lumen, may act as a midsystolic sphincter.

Although the side of re-entry has not been established, it is most likely that the false lumen encroaches upon the left common iliac artery to produce the differences in leg pulses, and it is reasonable to place the site of re-entry near the origin of this vessel.

Although the extent of aortic involvement in this man is as great as in Case 1, only one episode of pain occurred, and pain was not felt in the back or below the xiphoid. Much of the dissection process must have been painless, a not uncommon feature. It is also noteworthy that dissection of the innominate has not continued, despite untreated hypertension and the presence of the false lumen which might be expected to act as a wedge and overcome the natural cohesive forces of the vessel.

**Summary of haemodynamic findings** In both patients, sottography has confirmed flow of blood in both lumina. The false lumen extends from near the right coronary ostium to the aortic
bifurcation. Both have moderate hypertension, dilatation of the aortic root, and aortic incompetence. Both have a normal cardiac output at rest, and both respond to mild effort with no significant change in stroke volume but with an abnormal widening of the AV oxygen difference. Case 2 has the more severe hypertension and somewhat inferior left ventricular function.

**Discussion**

Dissecting haematoma of the aorta is by no means a new disease.

The earliest reports do not clearly distinguish between true aneurysms, traumatic partial ruptures (false aneurysms), and dissecting haematoma. The first undoubted description of the dissection process we owe to Maunoir (1802) — '...le sang disèque quelquefois l'artère dans toute sa circonférence, et elle se trouve dans le centre de l'anévrisme entièrement baignée dans le sang anévrismal.' Twenty years later, Shekelton (1822) described chronic dissecting aneurysm, noting the re-entry opening, the atheroma, and the new endothelium of the sac, and appreciating its function. He stated in effect that the blood, forcing on, made a new passage into the aorta because the intima and media were weaker than the adventitia, a view that is probably correct. In 1843, Peacock gave a well-reasoned account of the development of dissecting haematoma, stating that the re-entry produced an 'imperfect natural cure' and gave the diagnostic signs of the condition. The first correct diagnosis of dissection in life was made by Swaine and Latham (1855). Despite this early start, by 1939 East could only find 27 reported cases diagnosed before death. The first surgical approach to dissection of the abdominal aorta (internal decompression) was reported by Gurin, Bulmer, and Derby (1935). DeBakey et al. (1955) pioneered surgery for both acute and chronic thoracic dissections. Today, it may be said that the pathogenesis and clinical features of acute dissection are well delineated. Treatment is primarily medical, with surgery in reserve for complications and failure of the medical regimen. The natural history and the prognosis of those who survive the first 6 months is less well known.

**Internal or external rupture** The initial lesion in this condition is a medial haematoma, gaining access to the lumen through a transverse intimal tear. Thereafter the haemorrhage shears the vessel wall into two coats of varying thickness by overcoming the natural cohesive forces, tending to proceed internally or externally depending upon the relative strength of the two layers. If the outer coat has the greater share of the collagen and elastic fibres, a second, deflating, intimal rupture may occur near a point of weakness due to atheroma. The site of rupture is also partly governed by the points of fixation of the aorta, notably the pericardial reflexion, the ligamentum arteriosum, the diaphragm,
dissection towards the aortic root must not produce catastrophic aortic regurgitation, occlude the coronary arteries, or enter the pericardium. Antegrade dissection must not occlude major branches, notably the brachiocephalic vessels, the intercostals and the spinal blood supply, the midline anterior visceral vessels, and the lateral arteries, particularly the renals. If the patient does not succumb to these complications or to shock, the process may either cease, allowing the haematoma slowly to resorb, fibrose, and heal, or it may re-enter by internal rupture. What then is the prognosis of a patient whose dissecting haematoma has been stabilized in this manner?

Prolonged survival Several examples of prolonged survival have been reported. In some, there was no clear-cut antemortem diagnosis. Once the condition had been discovered at necropsy, a search was made for the false lumen. The innominate artery is narrowed by the false lumen. Above this narrowing there is poststenotic dilatation of the innominate artery.

or the major branches, including the bifurcation.

Once internal rupture and decompression occurs, the total strength of the aortic wall is not necessarily greatly diminished. Since the pathway via the false lumen is longer and more tortuous than that through the true lumen, the peaks of pressure in the two lumens may be out of phase, so that the peak distending forces may be dispersed in time and area.

Experimentally, progression of dissection from an intimal tear is related to pulsation, in particular to the maximal rate of pressure rise, and does not occur with nonpulsatile pressures up to 400 mmHg (Prokop, Palmer, and Wheat, 1970). Therefore, formation of a double barrel aorta does not, per se, increase the probability of external rupture. Late rupture, in all probability, results from a recurrence of medial haemorrhage, i.e. from a fresh dissection.

Immediate survival For a patient with an acute dissecting haematoma of the aorta to survive the acute episode, without operation, several fortunate occurrences are necessary. The lesion must not penetrate the adventitia to produce external rupture. Retrograde
(1886) reported a 30-year survival, Böstrom (1888) one of 22 years, and Hall (1926) one of 15 years. Rineberg, Forline, and Orgain (1960) reported a 19-year survival in a case without angiographic or necropsy confirmation, but in which the diagnosis can hardly be doubted. Jones and Langley (1946) observed a case alive and well 8 years after the initial diagnosis was made: according to Rineberg et al. (1960) that diagnosis was confirmed at necropsy when the patient died of unrelated causes, 14 years after the acute dissection. Schatz (1969) described 2 cases of aortic dissection surviving 74 and 26 months: in both, progressive enlargement of the aneurysm was recorded, and both died suddenly.

Of patients with an undoubted time of onset and proof of the lesion, Case 1 of this paper is the longest survivor, at 18 years. One may conclude that a double-barrel aorta is not necessarily incompatible with prolonged survival and useful life.

Diagnosis of chronic dissecting haematoma Though the physical signs of acute dissection are well known, those of a chronic aortic haematoma are not. The diagnosis may, however, be strongly suspected if the significance of certain findings is appreciated. A history of antecedent chest pain is important, but not essential, as the initial acute episode is occasionally painless. Generally, mild to moderate hypertension is present, and aortic incompetence is frequent. Diminution and delay of the pulse in the right carotid and subclavian arteries and their branches indicate narrowing of the innominate artery. This is well shown in Case 2 (Fig. 6), and was also present in W. S. Middleton’s case (1970, personal communication). This sign is the reverse of what may be found in a pre-subclavian coarctation, where the left brachiocephalic pulsae may be diminished.

Diminution of one femoral pulse, or a unilateral Duroziez’ sign together with delay as compared to the left radial pulse, may also result from an aortic haematoma, suggesting that a common iliac artery is compressed.

The combination of such changes in the pulses with hypertension and aortic incompetence would be unusual in rheumatic or syphilitic aortic valve disease. When present, with or without a history of previous chest pain or ‘heart attack’, this triad should lead to a strong clinical suspicion of chronic aortic haematoma. The chest radiograph may be helpful, but retrograde aortography is essential to establish the diagnosis and allow consideration of rational therapy.

Mortality of chronic dissection Both Shennan (1934) and Hirst, Johns, and Kime (1958) have published extensive monographs on dissecting aortic haematoma. Their figures for causes of death in chronic cases vary somewhat. In the two series combined, 145 patients survived more than a few weeks. Of these, 53 (37%) died later of haemorrhage, 56 (39%) of cardiac failure, and 11 (8%) of renal failure. Advances in therapy since these classic reviews were written may significantly affect the late mortality experience.

Aortic incompetence is the main factor associated with the development of cardiac failure. It is usually due to distortion of the ascending aorta and dilatation of the annulus, and is amenable to surgical correction. Shennan considered hypertension the most important factor in determining the prognosis. Aortic rupture with haemorrhage is due to progression or recurrence of the dissecting process: the basis of Wheat’s treatment is the reduction of the force of cardiac impulse with each systole. Control of hypertension, and reduction of systolic force with propranolol, may therefore be of great value. Renal failure may be due to involvement of the renal arteries: Wheat considers renal arterial involvement an indication for operation. Recovery from a second acute dissection, some time after the first, is rare – only 8 cases having been reported (Hunter, 1963). Of these, 5 died from further dissection or rupture. Recurrence therefore suggests that the basic pathological processes are still present, and are widespread.

Arrested aortic haematoma Less common than healing by spontaneous internal decompression is the process which we may term arrest of the dissection. Further separation of the aortic coats does not take place. The blood in the lesion clots, to be replaced by fibrous tissue. This is equivalent to a healed partial rupture of the aorta (a false aneurysm). The incidence and prognosis of this form is quite unclear, though one may suspect that many medically treated cases will fall into this category. Kellaway (1963) reported a case in which healing was apparently complete at necropsy, the patient succumbing to the side-effects of treatment. In Middleton’s case (1970, personal communication) a physician is well 15 years after suffering an arrested haematoma involving the ascending aorta and innominate artery, the radiological shadow gradually diminishing over the years.

Therapy From the above it may be concluded that those who harbour a chronic
Aortic haematoma are not necessarily in dire straits, but can be managed rationally. Aortography is essential. If blood does flow in the false channel, treatment may be mainly conservative, with control of hypertension and propranolol. An operation may be considered for recurrence of acute dissection, for cardiac failure resulting from aortic regurgitation, for progressive enlargement of the sac, and for occlusion of the renal and other major arteries. On the other hand, if the false lumen does not carry blood, natural decompression has not taken place. If fibrosis and scar formation are going on, the radiographic shadow should show a continuous, if slow, shrinkage. If not, the lesion may well be unstable, and elective surgery may be wise.

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