Br Heart J 1982; 47: 294–7

Isolated atresia of the aortic arch in a 65-year-old man
Surgical treatment and review of published reports

SIMCHA MILO, CARLO MASSINI,* DANIEL A GOOR

From the Department of Thoracic and Cardiovascular Surgery and the Sheba Heart Institute, The Chaim Sheba Medical Center, Tel-Hashomer, Israel

SUMMARY A 65-year-old patient with isolated aortic arch atresia documented by cardiac catheterisation and cineangiography is described. Successful surgical repair was performed using a 10 mm Dacron tube graft to bypass the atretic segment.

Isolated atresia of the aortic arch is rare. In aortic arch atresia the arch is continuous, yet a segment of it is atretic so that apart from a collateral circulation, communication with the lower aortic segment is absent. Haemodynamically, the condition is similar to complete interruption of the aortic arch.

To our knowledge only 10 cases of isolated aortic arch atresia or interruption have been reported.[1-9] In eight cases, the diagnosis was confirmed at operation, one case was diagnosed but untreated,[8] and in one patient, the diagnosis was made at necropsy.[1]

In this report we present a 65-year-old man with isolated atresia of the aortic arch which was subsequently corrected by operation. The anatomical aspects, diagnostic problems, and surgical approach are discussed, with a review of the previously published cases.

Case history

A 65-year-old Bedouin man was referred to the Sheba Medical Centre for evaluation of shortness of breath, dizziness, palpitation, fatigue, and cachexia of one year's duration. On admission he was in poor general condition, cachectic, and dyspnoeic at rest. The blood pressure in the right arm was 145/120 mmHg but it could not be measured in the left arm. There was no cyanosis. Examination of the neck showed distinct enlargement of the neck veins. The right carotid artery was normal; the left carotid was palpable but very weak. The chest was symmetrical and aeration of the lungs was equal. The apex of the heart was visible and palpable in the sixth intercostal space in the anterior axillary line. Examination of the back disclosed arterial pulsation in the intercostal spaces on the right side only. The liver was enlarged 3 cm below the right costal margin and was tender to palpation. The femoral and peripheral leg pulses were not palpable bilaterally. The electrocardiogram showed sinus rhythm, enlargement of both atria, left ventricular hypertrophy, and strain. Chest x-ray film showed right sided rib notching. The haemoglobin was 14·7 mg/100 ml; WBC 7900; blood urea 7·0 mmol/l (42 mg/100 ml); serum creatinine 79·6 mmol/l (0·9 mg/100 ml). Cardiac catheterisation was performed via a percutaneous Seldinger right axillary approach. Thoracic aortography showed interruption of the aortic arch before the origin of the left subclavian artery (Fig. 1A and B), with late filling of the descending thoracic aorta via a collateral circulation, especially the left vertebral artery (Fig. 2A and B).

At operation a left thoracotomy was made in the fourth left intercostal space. The aorta was dissected retrogradely, freeing the left subclavian artery which was small, approximately 3 mm in diameter. Further dissection of the aortic arch disclosed a narrow, 3 mm long segment of the aorta between the left subclavian artery and the left common carotid artery; no lumen was present. The left common carotid and right brachiocephalic trunk were dissected. A 10 mm woven Dacron graft was anastomosed to the left side of the brachiocephalic trunk using partial occlusion and was connected to the distal descending thoracic aorta using the same technique (Fig. 3). After completion of the anastomoses, a mean blood pressure of 80

*A visiting fellow from the Department of Cardiovascular Surgery, Ospedale Specializzato Regionale "G.M. Lancisi", Ancona, Italy.

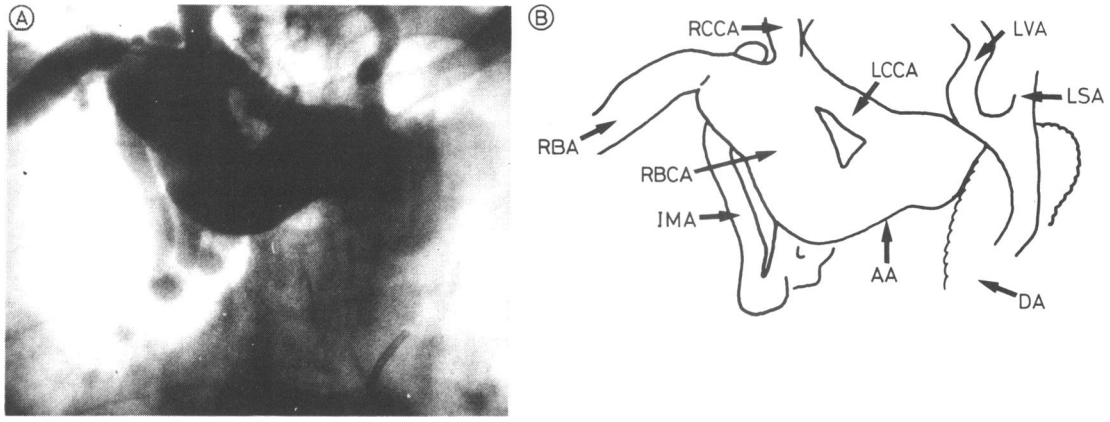

Fig. 1 *(A) The early phase of the ascending thoracic arteriogram in the right anterior oblique position. The brachiocephalic trunk is conspicuously dilated. Contrast material does not enter the descending thoracic aorta. (B) Diagrammatic representation of Fig. 1A. Contrast material introduced into the ascending aorta (AA) through the right brachial artery (RBA) flows into the right brachiocephalic artery (RBCA) and right common carotid artery (RCCA), left common carotid artery (LCCA), and the internal mammary artery (IMA). The descending aorta (DA) is supplied later through the vertebrobasilar system. Blood flows via the left vertebral artery (LVA) into the left subclavian artery (LSA) and into the descending aorta.*

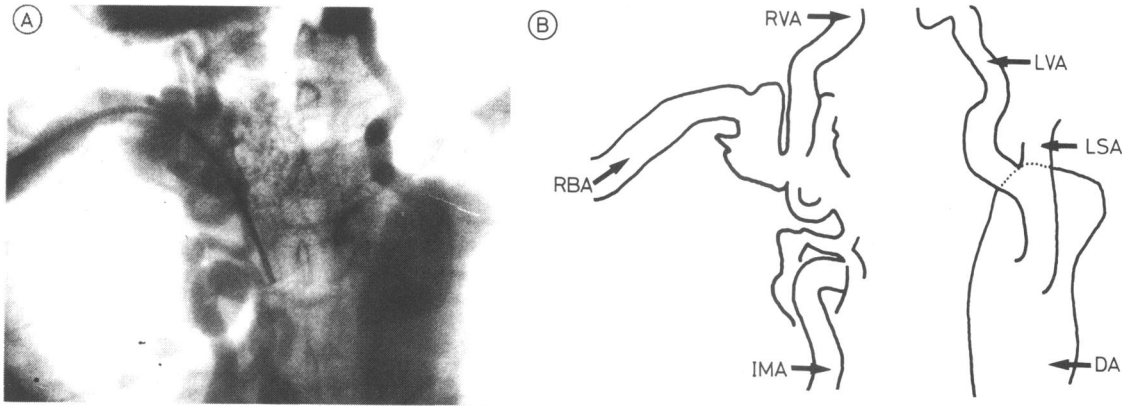

Fig. 2 *(A) A late phase picture of the thoracic arteriogram in the right anterior oblique position. The internal mammary artery is much enlarged and tortuous. The descending thoracic aorta is now visualised. The right and left vertebral arteries are seen with dense opacification of the left vertebral artery. (B) Diagrammatic representation of Fig. 2A. RBA, right brachial artery; RVA, right vertebral artery; LVA, left vertebral artery; LSA, left subclavian artery; DA, descending thoracic aorta; IMA, internal mammary artery.*

mmHg was measured in the ascending aorta and a mean pressure of 70 mmHg in the descending aorta. Both femoral pulses and the left radial pulse were palpable and of good volume. Twenty-four hours after operation simultaneous pressure measurements were made in the right brachial artery and right femoral artery. Systolic pressure in the right brachial artery was 120 mmHg and in the femoral artery 108 mmHg.

On discharge the right arm blood pressure was 130/90 mmHg. Nine months after operation the patient was well but has subsequently left the country and has not been restudied radiologically.

Discussion

Atresia of the aortic arch is a rare malformation and has been reported mainly in association with other cardiac anomalies such as persistent ductus arteriosus and ventricular septal defect. The anomaly is different anatomically from interruption of the aortic arch,

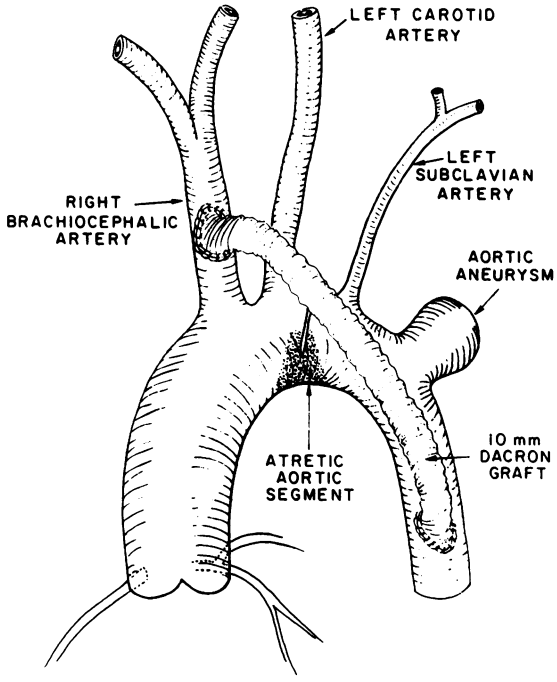

Fig. 3 *Pathological anatomical findings in our patient as seen during operation and the operative treatment. The atretic segment was found between the left common carotid artery and the small left subclavian artery.*

although haemodynamically similar. Whereas in interruption of the aortic arch part of the arch is absent completely, with varying distance between the two ends of the aorta, in atresia of the aortic arch the segment is short, externally narrow, and the lumen not patent. Only one previous case of aortic atresia without additional cardiac anomalies has been reported[6] while others were aortic arch interruptions[1-5 7 8] (Table).

Interruption of the aortic arch was first described in the eighteenth century by Steidele.[10] The currently adopted classification[11] divides the lesion into three types according to the site of interruption: A, interruption distal to the left subclavian artery; B, interruption between the left carotid and left subclavian arteries; C, interruption proximal to the origin of the left carotid artery.

The classification has also been used in atresia of the arch.[12]

In a series of 184 cases of aortic interruption,[13] 42% of the known and classifiable cases (70 of 165) were of type A, 53% of type B, and 4% type C. Almost all cases had associated anomalies: persistent ductus, ventricular septal defects, aorticopulmonary window, bicuspid aortic valve, and muscular subaortic stenosis. When associated with other cardiac malformation, the anomaly carries a 76% mortality within the first month of life,[14] the usual cause of death being early congestive heart failure and/or pulmonary complications. Interruption of the aortic arch with no other cardiac anomalies was thought to be incompatible with life[14] until Pillsbury and associates[2] reported, in 1964, a case in a 16-year-old girl who was operated on successfully. Our case is, to our knowledge, the oldest patient diagnosed and operated on, and is a case of isolated atresia of aortic arch, type B.

It is interesting that all but two reported patients with isolated aortic arch atresia carried their pathology far into adult life, in contrast to the large group of patients with additional intracardiac anomalies who

Table *Clinical data of 11 cases of isolated interruption and atresia of aortic arch*

Author	*Age (yr) and sex*	*Type*	*Blood pressure arms (mmHg)*		*Rib notching*	*Surgical approach*	*Operation*
			Right	*Left*			
Evans[1]	32 M	A (interrupted)	—	—	—	No surgery	
Pillsbury *et al.*[2]	16 F	C (interrupted)	140/100	—	Right side	Left thoracotomy	Bypass graft ♂ 10 mm Dacron tube
Asano *et al.*[3]	12 F	B (interrupted)	210/70	120/60	Right 9th rib	Left lateral thoracotomy, median sternotomy	Bypass graft ♂ 12 mm Dacron tube
Zetterqvist[4]	8 M	B (interrupted)	—	—	No notching	Unknown	Direct anastomosis between ascending and descending aorta
Morgan *et al.*[5]	19 M	B (interrupted)	95/70	95/70	No notching	Left thoracotomy	Bypass graft ♂ 10 mm Dacron tube
Le Page *et al.*[6]	56 M	A (atresia)	184/100	174/90	Right side	Unknown	Direct anastomosis
Kauff *et al.*[7] Case 1	36 M	B (interrupted)	180/120	140/110	Bilateral notching	Left thoracotomy	Direct anastomosis
Kauff *et al.*[7] Case 2	49 M	A (interrupted)	120/90	210/95	Bilateral notching	Left thoracotomy	Bypass ♂ Dacron tube graft
Judez *et al.*[8]	18 F	B (interrupted)	—	—	No notching	No surgery	
Sharratt *et al.*[9]	15 M	B (interrupted)	220/90	—	Right side	Median sternotomy and left thoracotomy	Bypass graft ♂ 20 mm Dacron tube
Present case	65 M	B (atresia)	145/120	—	Right side	Left thoracotomy	Bypass graft ♂ 10 mm Dacron tube

succumbed early in childhood.[14] Symptoms resembled those of long-standing systemic hypertension or aortic coarctation: headaches, fainting spells, visual disturbances, dysphasia, memory loss, dyspnoea, and leg pains on walking. Central nervous system symptoms seen in cases with interruption of types B and C are probably the result of vertebral steal.[4 9] Retrograde flow from the left vertebral and carotid arteries to the descending aorta was present in our patient and carotid arteriograms showed filling of the vertebrobasilar system from an injection into the right carotid artery (Fig. 2A and B). Reconstitution of blood flow to the descending aorta was achieved at operation, either by direct anastomosis (cases 4, 7, 9) or by use of a synthetic tube graft, depending on the anatomical distance between the two end pouches of the ascending and descending aorta and the feasibility of bringing them together. Operation was successful in all patients included in this review. Surgical treatment of the anomaly is relatively simple and produces distinct improvement in both neurological and cardiovascular status.

References

1 Evans W. Congenital stenosis (coarctation), atresia, and interruption of the aortic arch — (a study of twenty-eight cases). *Q J Med* (NS) 1933; **2**: 1–31.

2 Pillsbury RC, Lower RR, Shumway NE. Atresia of the aortic arch. *Circulation* 1964; **30**: 749–54.

3 Asano K, Shiozaki K, Eguchi S, Irisawa T. Congenital aortic arch interruption: report of a case, surgically treated with success. *Jpn Heart J* 1967; **8**: 196–201.

4 Zetterqvist P. Atypical coarctation of the aorta with bilateral vertebral-subclavian pathway. *Scand J Thorac Cardiovasc Surg* 1967; **1**: 68–75.

5 Morgan JR, Forker AD, Fosburg RG, Neugebauer MK, Rogers AK, Bemiller CR. Interruption of the aortic arch without a patent ductus arteriosus. *Circulation* 1970; **42**: 961–5.

6 Le Page JR, Viamonte M Jr, Jude JR. Complete interruption of the arch of the aorta: an enigma? *J Can Assoc Radiol* 1971; **22**: 60–4.

7 Kauff MD, Bloch J, Baltaxe HA. Complete interruption of the aortic arch in adults. *Radiology* 1973; **106**: 53–7.

8 Judez VM, Maitre MJ, de Artaza M, deMiguel JMF, Valles F, Marquez J. Interruption of aortic arch without associated cardiac abnormalities. *Br Heart J* 1974; **36**: 313–7.

9 Sharratt GP, Carson P, Sanderson JM. Complete interruption of aortic arch, without persistent ductus arteriosus, in an adult. *Br Heart J* 1975; **37**: 221–4.

10 Steidele RJ. *Sammlung verschiedener in der chirurgisch-praktischer Lehrschule gemachten Beobachtungen.* vol. 2. Vienna: Gräffer, 1777–1778: 114.

11 Celoria GC, Patton RB Congenital absence of the aortic arch. *Am Heart J* 1959; **58**: 406–13.

12 Tawes RL Jr, Panagopoulos P, Aberdeen E, Waterston DJ, Bonham-Carter RE. Aortic arch atresia and interruption of the aortic arch. Experience in 11 cases of operation. *J Thorac Cardiovasc Surg* 1969; **58**: 492–501.

13 Van Praagh R, Bernhard WF, Rosenthal A, Parisi LF, Fyler DC. Interrupted aortic arch: surgical treatment. *Am J Cardiol* 1971; **27**: 200–11.

14 Roberts WC, Morrow AG, Braunwald E. Complete interruption of the aortic arch. *Circulation* 1962; **26**: 37–59.

Requests for reprints to Dr Simcha Milo, Department of Thoracic and Cardiovascular Surgery, The Chaim Sheba Medical Center, Tel-Hashomer, Israel.