Coarctation of the aorta in monozygotic twins

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SUMMARY Concordant coarctation of the aorta has been found in a pair of monozygotic twins. One of the twins had a vascular ring.

The aetiology of coarctation of the aorta is still largely unexplained, and investigations on familial occurrence have given conflicting results.¹⁻³

Only 10 reports of coarctation of the aorta in monozygotic twins have been published. The lesion has shown discordance^{1,4-9} in all but one¹⁰ of the cases. In only three of these was there a concordant cardiovascular malformation: atrial septal defect,⁵ hypoplasia of the aortic arch,⁸ and Marfan's syndrome,⁹ together with coarctation of the aorta in one of the twins.

In this communication we describe coarctation of the aorta in a monozygotic pair of twins: the second reported case with concordance, and the first case where coarctation was found alone in one of the twins and appeared with a vascular ring in the other.

Case report

From the time of birth (May 1963) the twin girls were regarded as being monozygotic. Sixteen years later this assumption was confirmed by blood type, plasma enzyme and tissue type investigations, and by chromosome marker investigations, using a fluorescent banding method (see Table).

The vascular anomalies were assessed by means of cardiac catheterisation and angiocardiology as well as by direct inspection of the thoracic aorta during operation.

The twin with coarctation and vascular ring malformation, twin S, measured 49 cm at birth and had surgery at the age of almost 1 year. At operation the trachea was decompressed by division of both the

ligamentum arteriosum and the totally coarctated segment of the pretracheal anterior aortic arch. The post-tracheal segment of the vascular ring had the size and function of a normal aortic arch, continuing into the descending aorta. The patient has since developed normally.

The twin with the isolated coarctation, twin J, measured 44 cm at birth and had an operation when 16 years old. The coarctation was of the so-called postductal type and was treated with a bypass graft.

Both girls were seen three months after twin J's operation. The respective heights were 152.5 cm for S and 147.5 cm for J. After 20 minutes' rest the blood pressures taken on the right arm were, respectively, 135/66 mmHg for S and 136/64 mmHg for J, who had a pressure of 145/80 mmHg preoperatively.

Ophthalmological examination showed that J's retinal arteries were more tortuous than those of S.

Discussion

In our department, in addition to the two cases reported here, we have operated on 232 patients with coarctation of the aorta. Four of these came from two families, a boy and his aunt and a brother and sister. This observation only lends limited support to the suggestion of Simon *et al.*³ that there is "a genetic contribution to the aetiology of this disorder".

It has long been accepted that overdevelopment of the upper part of the body is characteristic of coarctation of the aorta. One of our twins (J) had had her coarctation and high proximal blood pressure for 16 years and yet showed no difference in the build of the

Table Genetic evidence of zygosity

	ABO	Rhesus C c	D	E	e	MNS	PI	Lu	K	Le ab	Fy ab	Jk ab	Bf	C2	Glo	HLA
Twin J:	A2	-	+	+	+	+++	+	a-	-	++	++	+-	FS	1	1	A2, B7/A9, Bw39
Twin S:	A2	-	+	+	+	+++	+	a-	-	++	++	+-	FS	1	1	A2, B7/A9, Bw39

The twins had identical QM banded karyotypes. Informative markers were found in the following chromosome numbers: 1, 3, 4, 9, 13, 14, 15, 16, 21, and 22.

trunk, neck, or head compared with the other. Furthermore, twin J had shorter lower limbs accounting for the difference in height between the two. The same observation has been made by Driver and Suckling.⁵ This might indicate that the characteristic build in this condition is caused more by underdevelopment of the lower half of the body than by overdevelopment of the upper half.

The investigation on blood types, and enzyme and tissue types was performed at the Blood Bank and Tissue Typing Laboratory, Aarhus Kommunehospital; the chromosome analysis was done at the Institute of Human Genetics, Aarhus University, Aarhus, Denmark. I am grateful to both these laboratories for their collaboration.

References

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Notices

Subacute bacterial endocarditis

A survey is currently being carried out by the British Cardiac Society and the Medical Services Study Group of the Royal College of Physicians. Though improvement of dental prophylaxis is one objective, the survey is already yielding other valuable information. It is hoped that proformas will be received in respect of a high proportion of patients with subacute bacterial endocarditis in the British Isles seen during 1981 and 1982 and readers are asked to arrange for them to be submitted in respect of any cases that come to their notice. Proformas can be obtained from Sir Cyril Clarke, Medical Services Study Group, King's Fund Centre, 126 Albert Street, London NW1 7NF (tel. 01-267 6111, ext. 263) to whom they should be returned.

British Cardiac Society

The Autumn meeting will take place at Wembley on 6 and 7 December 1982 and the closing date for receipt of abstracts is 11 August 1982.

The Annual General Meeting for 1983 will take place in Bristol on 12 and 13 April, and the closing date for abstracts will be 4 January 1983. The Autumn Meeting will be held at Wembley on 21 and 22 November 1983, and the closing date for abstracts will be 28 July 1983.