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Distinguishing between anomalous origin of the left coronary artery from the pulmonary trunk and dilated cardiomyopathy: role of echocardiographic measurement of the right coronary artery diameter

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SUMMARY Patients with anomalous origin of the left coronary artery from the pulmonary trunk usually have a large right coronary artery. This study examines the diagnostic value of measuring the diameter of the right coronary artery by echocardiography in distinguishing between this lesion and other causes of dilated cardiomyopathy. The diameter of the right coronary artery and the right coronary artery/aorta ratio were measured in the parasternal short axis view in 40 controls, 11 patients with dilated cardiomyopathy, and 10 with anomalous origin of the left coronary artery from the pulmonary trunk. In the controls, the diameter of the right coronary artery increased with age, but the right coronary artery/aorta ratio remained constant. In the control group the 95% upper limits of prediction for right coronary artery diameter were 1·6 mm for one month of age, 1·8 mm for three months, 2·0 mm for one year, 2·2 mm for two years, 2·4 mm for three years, 2·6 mm for four years, 2·7 mm for six years, 3·0 mm for eight years, and 3·2 mm for 10 years; and for right coronary/aorta ratios the limits were 0·17 for one month to one year, 0·18 for one to six years, 0·19 for six to 10 years, and 0·20 for more than 10 years. All patients with dilated cardiomyopathy had normal right coronary artery diameters and right coronary artery/aorta ratios (0·10–0·13). Those patients with anomalous origin of the left coronary artery from the pulmonary trunk had larger than normal right coronary artery diameter and a significant increase in the right coronary artery/aorta ratio (0·21–0·29).

The presence of an anomalous left coronary artery was likely if the diameter of the right coronary artery or the right coronary artery/aorta ratio was larger than the normal 95% limits of prediction.

Anomalous origin of the left coronary artery from the pulmonary trunk is one of the rare causes of dilated cardiomyopathy in infants and children.[1] Because an operation may give better results than medical treatment, early recognition and differentiation from other causes of dilated cardiomyopathy may be important in preventing the development of extensive myocardial damage.[2–4] Recent advances in cross sectional echocardiography permit the diagnosis to be made non-invasively[5]; however, it may not always be possible to image the origin of the left coronary artery from the pulmonary trunk.[6] Similarly, confusion with the transverse sinus has given rise to false positives.[7] On the other hand, the right coronary artery is usually dilated and tortuous in anomolous origin of the left coronary artery from the pulmonary trunk.[8,9] This report assesses the role of measurement of diameter of the main right coronary artery by cross sectional echocardiography in distinguishing between this lesion and other forms of dilated cardiomyopathy.

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Patients and methods

We studied 10 patients with anomalous origin of the left coronary artery from the pulmonary trunk (aged 3–79 months, median 7·5 months) (table 1), 11 with

Table 1 *Patients with anomalous origin of the left coronary artery from the pulmonary trunk*

No	*Age (mnth)*	*RCA (mm)*	*AO (mm)*	*RCA/AO*
1	3	2·6	9·0	0·29
2	3	2·2	9·0	0·26
3	4	2·3	9·5	0·24
4	7	2·2	10·0	0·22
5	8	2·0	10·0	0·20
6	9	3·0	11·0	0·27
7	15	3·5	12·0	0·29
8	55	3·6	17·5	0·21
9	70	4·4	15·5	0·28
10	79	3·9	18·0	0·22

AO, diameter of the aortic root; RCA, diameter of the right coronary artery; RCA/AO, ratio of right coronary artery to aorta.

Table 2 *Patients with dilated cardiomyopathy*

No	*Age (mnth)*	*RCA (mm)*	*AO (mm)*	*RCA/AO*	*Aetiology*
1	2	0·9	9·0	0·10	EFE
2	3	1·1	9·0	0·12	EFE
3	4	1·1	9·5	0·12	EFE
4	6	1·1	10·0	0·11	EFE
5	6	1·2	10·0	0·12	EFE
6	7	1·2	10·0	0·12	EFE
7	7	1·1	10·0	0·11	EFE
8	13	1·2	12·0	0·10	EFE
9	44	1·7	13·0	0·13	ICM
10	54	1·6	14·0	0·11	ICM
11	113	1·9	17·0	0·11	ICM

AO, diameter of the aortic root; EFE, endocardial fibroelastosis; ICM, idiopathic cardiomyopathy; RCA, diameter of the right coronary artery; RCA/AO, ratio of right coronary artery to aorta.

dilated cardiomyopathy of another cause (aged 2–113 months, median 7 months) (table 2), and 40 controls (aged 1–123 months, median 29·5 months). The cause of the dilated cardiomyopathy was endocardial fibroelastosis in eight patients and idiopathic cardiomyopathy in three patients.

We reviewed videotapes recorded from a standard approach with a 7·5 MHz transducer. We used a parasternal short axis view at the level of the right coronary artery for all patients and controls. The vessel was measured between 0·5 and 1 cm from the ostium. This enabled the diameter to be assessed in

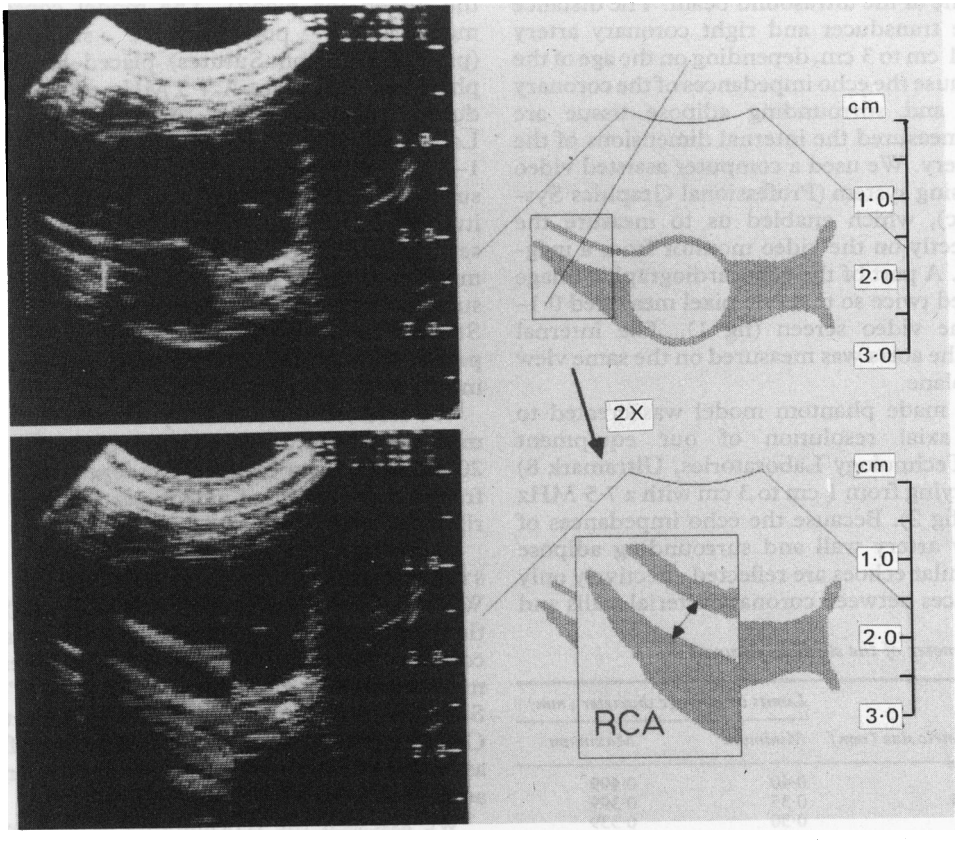

Fig 1 *Echocardiographic method for measurement of right coronary artery (RCA) and aortic diameters. Measurement line is aligned with the axial beam. Right coronary artery is measured in a magnified image by PGS (Professional Graphic System, Symtec).*

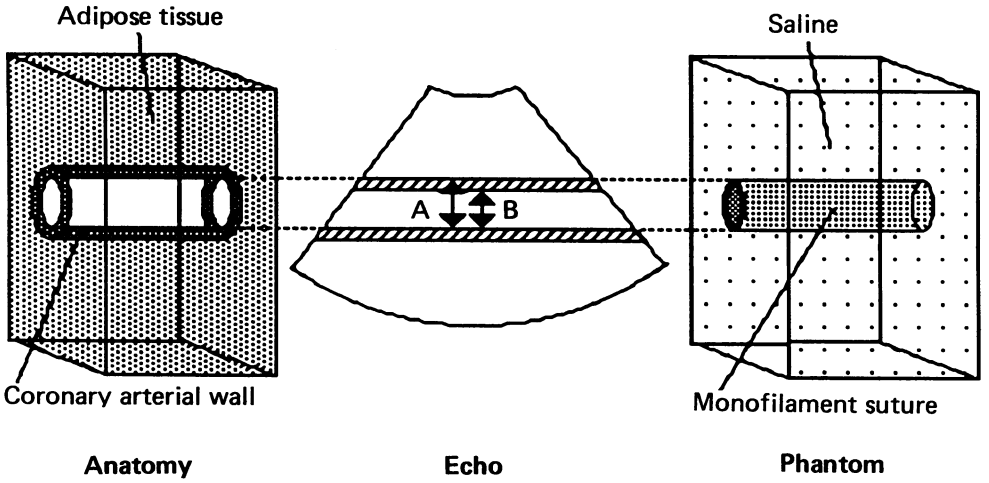

Fig 2 *Custom phantom model for assessing the accuracy of the measurement of the coronary artery. A, leading edge method; B, internal measurement. See text for explanation.*

the axial plane of the ultrasound beam. The distance between the transducer and right coronary artery varied from 1 cm to 3 cm, depending on the age of the patient. Because the echo impedances of the coronary artery wall and surrounding adipose tissue are similar, we measured the internal dimensions of the coronary artery. We used a computer assisted video image digitising system (Professional Graphics System, Symtec), which enabled us to measure the distance directly on the video monitor from a magnified image. A part of the echocardiographic image was magnified twice so that one pixel measured 0·1–0·3 m on the video screen (fig 1). The internal diameter of the aorta was measured on the same view in the axial plane.

A custom made phantom model was created to assess the axial resolution of our equipment (Advanced Technology Laboratories, Ultramark 8) at depths varying from 1 cm to 3 cm with a 7·5 MHz transducer (fig 2). Because the echo impedances of the coronary artery wall and surrounding adipose tissue are similar echoes are reflected effectively only at the interfaces between coronary arterial walls and the lumen (or blood). The model consisted of a monofilament polypropylene surgical suture (prolene, Ethicon Sutures) placed horizontally in physiological saline. A 7·5 MHz ultrasound transducer (Access C, Advanced Technology Laboratories, 6·4 mm in diameter, 2 cm focus point, 1–4 cm focus zone) was placed 1–3 cm above the suture. The cross sectional image showed the longitudinal axis of the suture. We used minimal gain settings to exclude reverberation echoes. We determined the limits of axial resolution by examining sutures that varied in size from 1 to 4–0 USP (United States Pharmacopeia) (table 3). Two horizontal parallel lines were derived from the upper and lower interfaces of the suture.

Table 3 *Diameter of the surgical sutures**

		Limits on average diameter (mm)	
USP size	*Metric size (mm)*	*Minimum*	*Maximum*
1	4	0·40	0·499
0	3·5	0·35	0·399
2–0	3	0·30	0·339
3–0	2	0·20	0·249
4–0	1·5	0·15	0·199

**United States Pharmacopeia XXI*, p 1009.

We assessed the reliability of the vessel diameter measurement by cross sectional echocardiography in 20 controls. Two observers independently selected frames from recorded videotapes and measured the right coronary artery and aorta twice.

STATISTICAL ANALYSIS

We calculated a regression line and the 95% prediction limits of the right coronary artery and right coronary artery/aorta ratio with the general linear models (GLM) procedure in SAS User's Guide: Statistics (Version 5 Edition, SAS Institute, North Carolina, 1985). Student's *t* tests were performed to assess the differences in the right coronary artery/aorta ratio between groups.

We assessed the repeatability of a measurement from the means and standard deviations calculated for intra-investigator and inter-investigator differences.

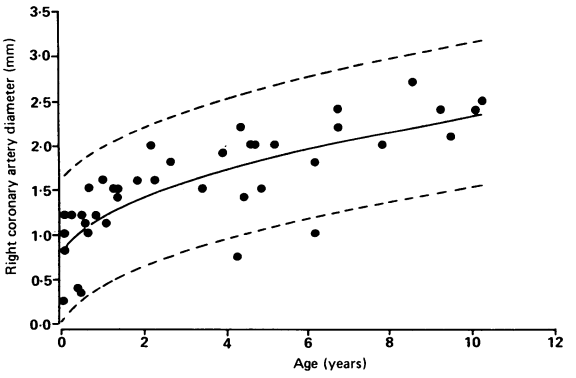

Fig 3 *Normal right coronary artery diameters with age by echocardiographic measurement. The regression equation line and 95% prediction limits are given.*

Results

RESOLUTION BY PHANTOM MODEL STUDY

When we used the zoom function of the equipment we were able to resolve two parallel lines of a USP size 2–0 suture (0·30–0·339 mm in diameter) with an absolute echo free space between the lines, but we could not resolve two lines for a USP size 0 suture (0·20–0·249 mm in diameter). Therefore the resolution of this system must lie between 0·2 and 0·34 mm. Theoretically, the leading edge method (arrow line A in fig 2) is more accurate for the assessment of vessel diameter; however, in the clinical setting scattered echoes from the surrounding tissue interfere with clear identification of the upper leading edge. For this reason we used the internal dimension of the coronary artery image (arrow line B in fig 2). This method therefore must underestimate the true diameter by 0·2–0·34 mm.

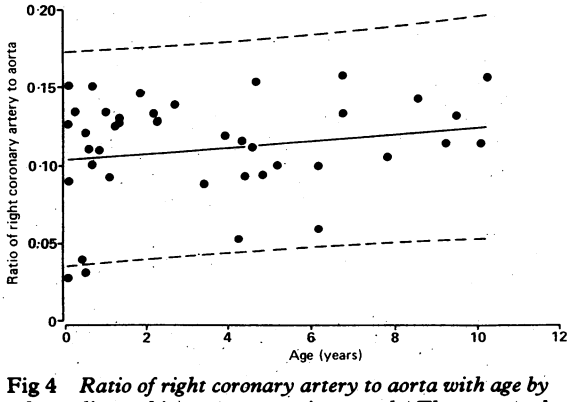

Fig 4 *Ratio of right coronary artery to aorta with age by echocardiographic measurement in controls. The mean and 95% prediction limits are given.*

Table 4 *Normal values of right coronary artery diameter and ratio of right coronary artery to aorta with age*

	RCA (mm)		*RCA/AO*	
Age	*Mean*	*95% upper limit*	*Mean*	*95% upper limit*
1 mnth	0·8	1·6	0·11	0·17
2 mnth	0·9	1·7	0·11	0·17
3 mnth	1·0	1·8	0·11	0·17
6 mnth	1·1	1·9	0·11	0·17
1 yr	1·2	2·0	0·12	0·18
2 yr	1·4	2·2	0·12	0·18
3 yr	1·6	2·4	0·12	0·18
4 yr	1·8	2·6	0·12	0·18
6 yr	1·9	2·7	0·12	0·19
8 yr	2·2	3·0	0·12	0·19
10 yr	2·5	3·2	0·13	0·20

NORMAL RIGHT CORONARY ARTERY DIAMETER

Several linear regression models were fitted for the effect of age and various transformations of age (including log and square root of age) on right coronary artery diameter. The best fit was that between the square root of age and right coronary artery diameter ($p = 0{\cdot}0001$. $R^2 = 0{\cdot}62$) (fig 3).

Right coronary artery (mm) = $0{\cdot}692 + 0{\cdot}151\sqrt{}$(age in months), standard error for the estimated slope coefficient = 0·019

Figure 3 gives prediction limits for the normal right coronary arteries. There was no evidence of an age effect on the right coronary artery/aorta ratio ($p = 0{\cdot}21$, $R^2 = 0{\cdot}04$) (fig 4): right coronary artery/aorta ratio = $0{\cdot}104 + 0{\cdot}00018$ (age in months), standard error for the estimated slope coefficient = 0·00014. Table 4 lists the calculated means and 95% upper limits of prediction for right coronary artery diameter and right coronary artery/aorta ratio for individual ages.

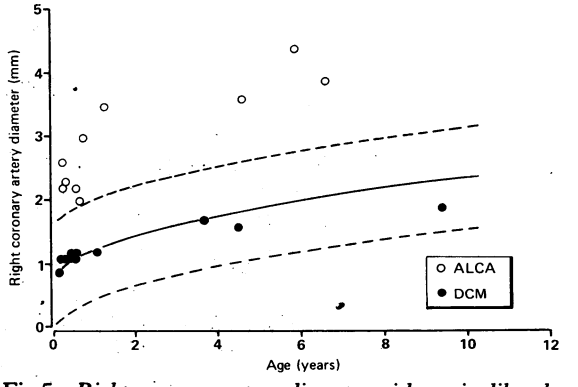

Fig 5 *Right coronary artery diameter with age in dilated cardiomyopathy (DCM) and anomalous origin of the left coronary artery from the pulmonary trunk (ALCA) by echocardiographic measurement. The 95% prediction limits of normal are given.*

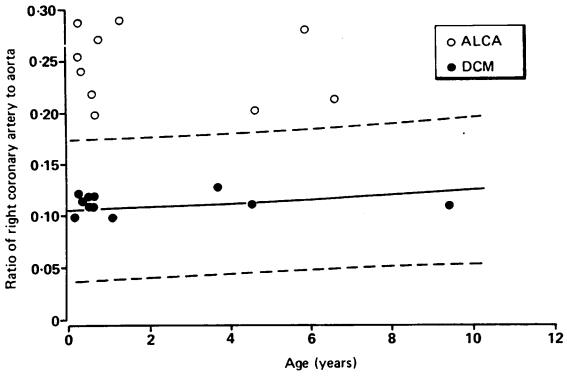

Fig 6 *Ratio of right coronary artery to aorta with age in dilated cardiomyopathy (DCM) and anomalous origin of the left coronary artery from the pulmonary trunk (ALCA) by echocardiographic measurement. The mean and 95% prediction limits of normal are given.*

DILATED CARDIOMYOPATHY

The right coronary artery and right coronary artery/aorta ratio were all within the normal range in patients with dilated cardiomyopathy (figs 5 and 6). The right coronary artery/aorta ratio ranged from 0·10–0·13 (mean (SD) 0·11 (0·009)).

ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY TRUNK

Direct imaging of the anomalous left coronary artery as it arose from the pulmonary trunk was only possible in five of 10 patients.

The size of the right coronary artery and the right coronary artery/aorta ratio were above the normal range in all patients with anomalous origin of the left coronary artery from the pulmonary trunk (figs 5 and 6). The right coronary artery/aorta ratio ranged from 0·21 to 0·29 with a mean (SD) 0·25 (0·034) that was significantly greater than that in controls and in patients with a dilated cardiomyopathy ($p < 0·001$).

INTRA AND INTER OBSERVER VARIABILITY OF MEASUREMENT

The mean difference between measurements made by the same investigator was 0·0183 (SD = 0·153, range −0·25 to 0·50, SE of a single observation 0·019 mm) for the right coronary artery, and 0·040 (SD 0·295, range −0·60 to −0·70, SE of a single observation 0·040 mm) for the aorta.

The mean difference between measurements by the two investigators was 0·044 (SD 0·148, range −0·18 to 0·40, SE of a single observation 0·024 mm) for the right coronary artery and −0·10 (SD 0·296, range −0·70 to 0·30, SE of a single observation 0·050 mm) for the aorta.

Discussion

DIFFICULTIES IN DISTINGUISHING BETWEEN ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY TRUNK AND OTHER FORMS OF DILATED CARDIOMYOPATHY

Anomalous origin of the left coronary artery from the pulmonary trunk in infants and children is often associated with a dilated cardiomyopathy because the collateral circulation from the right coronary artery is inadequate.[10 11] Early definitive diagnosis was based exclusively on aortography because clinical findings and ancillary investigations including chest *x* ray, electrocardiogram,[8 9] nuclear myocardial scintigraphy,[12] and analyses of left ventricular regional wall motion[13] did not distinguish this lesion from other forms of infantile dilated cardiomyopathy.

Fisher *et al* reported the first successful direct visualisation of the anomalous origin of the left coronary artery from the pulmonary trunk by cross sectional echocardiography in 1981.[5] This non-invasive test is much safer than aortography. This is a great advantage because anomalous origin of the left coronary artery from the pulmonary trunk is much rarer than other forms of dilated cardiomyopathy.[1] Although the sensitivity of direct imaging of the anomalous left coronary artery was thought to be adequate, false negative results have subsequently been reported.[6 7] In particular, it has been difficult to distinguish between the transverse sinus of the pericardium and the proximal portion of the left coronary artery. We believe that echocardiographic visualisation of the origin of the anomalous left coronary artery from the pulmonary trunk is still difficult even when the anomaly is strongly suspected because a child has a dilated poorly contractile left ventricle.

VALUE OF RIGHT CORONARY ARTERY VISUALISATION AND MEASUREMENT IN DIAGNOSING ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY TRUNK

The presence of a tortuous dilated right coronary artery in anomalous origin of the left coronary artery from the pulmonary trunk is well known.[8 9] It is apparent from this study that either a direct measurement of the right coronary artery or right coronary artery/aorta ratio can be used to distinguish between those patients with anomalous origin of the left coronary artery from the pulmonary trunk and other forms of dilated cardiomyopathy. Patients with anomalous origin of the left coronary artery from the pulmonary trunk always had values greater than the 95% upper limits of prediction. In our series the sensitivity and specificity was 100%.

This approach will miss only those rare cases who have pulmonary hypertension secondary to an associated intracardiac defect. These patients, however, should have normal left ventricular function, because coronary artery flow is preserved.[14 15] This technique will also identify patients with other diseases associated with a large right coronary artery—such as right coronary arteriovenous fistula, single right coronary artery, Kawasaki disease, or congenital cardiac anomalies with severe right ventricular hypertrophy. We hope that such cardiac causes will be excluded by clinical and echocardiographic examinations.

We are aware of the potential difficulties in obtaining accurate measurements of the right coronary artery diameter through echocardiographic imaging. With experience, however, the method is being successfully applied to normal children and those with Kawasaki disease.[16] At present we believe that all patients with an echocardiographic diagnosis of anomalous origin of the left coronary artery from the pulmonary trunk should have aortography.

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