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Original Article

Prevalence of asymptomatic coronary arterial abnormalities detected by angiography in grown-up patients with congenital heart disease

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Abstract *Aims:* Coronary arterial abnormalities, congenital and acquired, may co-exist with structural congenital malformations of the heart. This study analyses information obtained from coronary angiography in grown-up patients with congenital cardiac disease undergoing cardiac catheterisation. *Methods and Results:* We studied 448 patients, of whom 181 (40%) had selective coronary angiography. This was undertaken as a routine measure in 165 (37%), and because of clinical indications in 16 (4%). Aortic root angiography was carried out in 107 (24%). Coronary arterial abnormalities were demonstrated in one-third of those undergoing selective angiography, and in 69% of those with a clinical indication for angiography, compared with 30% who had a routine procedure. In those patients undergoing aortography alone, abnormalities were recognized in 17%, while the angiographic images were inconclusive in 24%. In the remaining 59%, no abnormality was apparent, but only the proximal coronary arteries could be seen clearly. In total, congenital abnormalities of the coronary arteries were found in 11% of the patients, acquired lesions were demonstrated in 5%, and surgically acquired lesions in 2%. *Conclusion:* Routine coronary angiography during catheterization of grown-up patients with congenital heart disease revealed a high incidence of abnormalities of the coronary arteries. Selective injection into the arteries had a higher sensitivity than aortography. Consideration should be given to performing routine coronary arteriography during invasive investigation of patients with congenital cardiac disease, as it provides useful and often unexpected information, which may be valuable for the interpretation of symptoms and the planning of subsequent surgery.

Keywords: Congenital cardiac disease; coronary angiography; coronary arterial abnormalities

WITH AN INCREASING NUMBER OF PATIENTS with congenital heart disease reaching adolescence and adulthood, a new population is presenting to adult physicians. These are patients referred with cardiac problems associated with complex structural anomalies following earlier surgical procedures.[1–5] Such patients may require invasive investigation to delineate haemodynamic problems.

The adult cardiologist without experience of congenital cardiac disease may be unfamiliar with the techniques required to obtain good haemodynamic and anatomical data, but is likely to be adept at producing high quality coronary arteriograms. In contrast, the paediatric cardiologist is used to producing good haemodynamic and anatomical data, but is frequently disinclined to attempt selective coronary arteriography, relying on an injection in the aortic root to show the origins of the coronary arteries.

Since congenital abnormalities of the course and origin of the coronary arteries, along with surgically acquired and atherosclerotic lesions, may occur in this ageing group of patients, routine coronary arteriography during invasive investigation may be useful. This study analyses the prevalence of asymptomatic abnormalities of the coronary arteries in such a group of patients, as well as the local practice of paediatric

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and adult cardiologists, during invasive investigation of patients admitted to a unit dedicated to the management of grown-up patients, including adolescents and adults, with congenital cardiac disease.

Material and methods

During the 5-year period of study, 461 patients aged 13 years and over, with congenital cardiac malformations, underwent cardiac catheterization that included study of the left heart. Of this number, 13 were subsequently excluded because their angiograms could not be found. The analysis was made, therefore, on 448 patients, 246 male and 202 female, of mean age 28.4 years, with a range from 13 to 76 years, and with a median age of 25 years. Data were collected by retrospective and prospective review of medical records and angiograms. The coronary arteriograms, and aortic root angiograms, were reviewed by two cardiologists, who reached a consensus on the origin, distribution and pathology of the coronary arteries. Abnormalities of the coronary arteries were diagnosed following the guidelines of Ishikawa and Brandt.[6] It was noted whether the investigation had been performed by a paediatric or adult cardiologist.

Statistical analysis

Groups were compared using the chi-squared test, and continuous variables with Student's *t*-test. A probability of the null hypothesis being correct of less than 0.05 was considered significant.

Results

The distribution of underlying cardiac diagnoses in the patient group is illustrated in Figure 1. Patients were divided into three groups according to the coronary imaging attempted at catheterisation. This division, along with the major findings involving the coronary arteries, is summarized in Figure 2. Selective coronary arteriography was performed in 181 of the 448 patients (40%). Of these, the study was incomplete in 7, in whom only one coronary arterial orifice had been intubated for selective angiography despite the other orifice being evident on aortography. Of the patients undergoing selective coronary angiography, 16 had specific clinical indications, namely chest pain in 14 patients, an abnormal exercise test in one patient, and chest pain coupled with an abnormal exercise test in the remaining patient. The other patients all underwent coronary arteriography as a routine. Coronary arterial abnormalities were demonstrated in 60 patients (33%). Such abnormalities were found in 11 of the 16 patients (69%) with a specific clinical indication for coronary arteriography. In contrast, abnormalities were found in only 49 of the

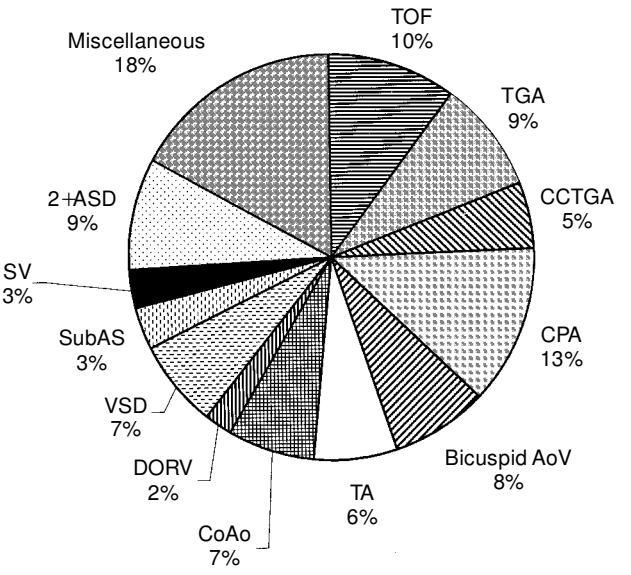

Figure 1.
Distribution of cardiac diagnoses within the study group. TOF: tetralogy of Fallot; TGA: complete transposition; CCTGA: congenitally corrected transposition; CPA: tetralogy of Fallot with complex pulmonary atresia; AoV: aortic valve; TA: tricuspid atresia; CoAo: coarctation of aorta; DORV: double outlet right ventricle; VSD: ventricular septal defect; SubAS: subaortic stenosis; SV: functionally single ventricle; 2° ASD: atrial septal defect within the oval fossa.

165 patients (30%) in whom arteriography had been performed as a routine measure ($p < 0.01$).

Aortic root angiography, without selective coronary arterial injection, was performed in 107 of the 448 patients (24%). The images proved inadequate for identification of lesions in 26 patients (24%). In the remaining 81 patients, abnormalities were identified in 18 (17%). In these patients, normal proximal arteries were seen in 63 (59%), but angiographic definition of the more distal vessels was poor.

No attempt had been made to image the coronary arteries in 160 patients (36%). During the period of study, 46 patients had one or more further cardiac catheterization, totaling 57 procedures. Of these, either selective coronary arteriography or aortography was performed in 24 patients who had not undergone specific imaging during their first procedure. Thus, in 136 (30%) patients, no attempt had been made to demonstrate the anatomy of the coronary arteries during the period of the study.

Paediatric cardiologists performed 219 of the catheterizations (49%), of which only 22 (10%) included routine or indicated coronary arteriography. Cardiologists from the adult congenital heart unit performed the remaining 229 (51%), of which 159 (69%) included coronary arteriography ($p < 0.001$). There was no obvious difference between the case mixes of these two groups. There was a trend towards an increasing rate of selective coronary arteriography

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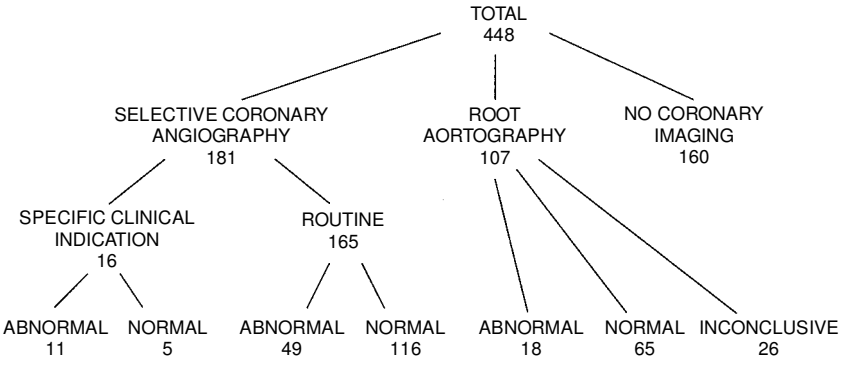

Figure 2.
Methods of coronary arterial imaging and diagnostic yield.

during the period of study, with 71 selective injections performed during the 245 procedures carried out over the first 30 months (29%) compared to 110 in 203 procedures (54%) during the second 30 months ($p < 0.001$).

In patients without specific indications for coronary arterial imaging, selective arteriography demonstrated abnormalities in 49 of 165 patients (30%), whereas aortography demonstrated abnormalities in 18 of 107 patients (17%). This difference was significant ($p < 0.05$).

Coronary arterial abnormalities

A total of 93 coronary arterial abnormalities were found in 78 patients, of whom 43 were male with a mean age of 35.0 years and a range from 14 to 70 years. The 35 female patients had a mean age of 33.6 years, and a range of 14 to 76 years. The mean age of all 448 patients studied was 26.0 years, with a range from 13 to 76 years. Congenital anomalies were found in 47 patients, while 24 had acquired disease, and 7 had lesions probably of iatrogenic origin, acquired at earlier reparative surgery. The type of abnormalities, and their distribution amongst the various cardiac diagnoses, is demonstrated in Table 1. The most frequent congenital abnormality was anomalous origin, seen in 25 patients, followed by a single coronary artery discovered in 12 patients with angiographic images of sufficient quality to exclude a second arterial orifice, fistulas or collateral arteries in 10 patients, separate origin of the left anterior descending and circumflex arteries in 8 patients, bifid origin of the right coronary artery in 7 patients, and hypoplasia of the anterior descending artery in 5 patients. Of patients with acquired disease, 17 had coronary atherosclerotic lesions, 6 patients had acquired coronary arterial collaterals in the setting of tetralogy of Fallot with complex pulmonary atresia, and one patient with coarctation of the aorta had an aneurysm of the left coronary artery. The mean age of patients with acquired coronary arterial disease was 48, with a standard deviation of ±17 years, this being significantly higher than those with congenital abnormalities, in whom the mean age was 29 years, with standard deviation of ±12 years ($p < 0.001$). The surgically acquired iatrogenic abnormalities included occlusion of a coronary artery in 4 patients, and stenosis in 3, with similar distributions between the left and right coronary arteries.

Discussion

Failure to recognize anomalies of the coronary arteries prior to surgical intervention may lead to accidental incision or ligation of the anomalous arteries. There are specific surgical pitfalls in certain situations, for example compromise of perfusion of myocardium supplied by a coronary artery with an anomalous origin during cardiopulmonary bypass,[2,5,7] ostial obstruction or more distal compression of an anomalous artery by a valvar prosthesis,[8,9] and the changes to perfusion pressure and oxygen saturation within an anomalous left coronary artery arising from the pulmonary trunk following radical repair of complex lesions. Thus, documentation of coronary arterial anatomy may be important in order to determine appropriate management in this group of adult patients, particularly if further cardiac surgery is required.

Our results highlight the fact that paediatric cardiologists are less likely to attempt selective coronary arteriography than adult cardiologists (10% vs. 69%, $p < 0.001$), while demonstrating that selective coronary arteriography is more sensitive at detecting abnormalities than is aortography alone (30% vs. 17%, $p < 0.05$). Previous studies have shown that selective arteriography gives a more accurate anatomical definition of the origins and distribution of the coronary arteries than aortography[10,11] or echocardiography,[12,13] these latter techniques showing only the proximal segments of the coronary vessels. Unorthodox projections during aortography, such as the 'end-on' or 'laid back' projection, specifically left anterior oblique projection with steep caudal angulation, have been shown to improve the sensitivity of aortography in the identification of coronary

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Table 1. Distribution of coronary arterial abnormalities by diagnosis.

Diagnosis	Abnormal coronary origin	Separate LAD & Cx origin	Hypoplastic LAD	Single CA	Anomalous course	CA fistula/ collaterals	Bifid RCA	Occluded/ stenosed CA	CA aneurysm	Atherosclerotic CA
TOF n = 9/46 (20%)	6		1	1				2		
TGA n = 7/40 (18%)	4	2	1	3	1		1			
CCTGA n = 6/23 (26%)	2	3	1				2			
TOF&CPA n = 7/58 (12%)	2		1			4	1			
Bicuspid AoV n = 6/35 (16%)	2					1		1		2
TA n = 5/29 (17%)	1			2		2				
CoAo n = 8/31 (26%)	2	1				2		1	1	2
DORV n = 4/11 (36%)	3			3						
VSD n = 6/31 (19%)		1		1			3			2
SubAS n = 3/15 (20%)	1	1								1
FSV n = 3/13 (23%)	1		1	1						
2° ASD n = 7/40 (18%)						1				6
Miscellaneous n = 9	1			1				3		4
TOTAL	25	8	5	12	1	10	7	7	1	17

Abbreviations: LAD: left anterior descending artery; Cx: circumflex artery; CA: coronary artery; PA: pulmonary artery; TOF: tetralogy of Fallot; TGA: concordant atrioventricular and discordant ventriculo-arterial connections; CCTGA: discordant atrioventricular and ventriculo-arterial connections; TOF&CPA: tetralogy of Fallot with complex pulmonary atresia; AoV: aortic valve; TA: tricuspid atresia; CoAo: coarctation of aorta; DORV: double outlet right ventricle; VSD: ventricular septal defect; SubAS: subaortic stenosis; FSV: functionally single ventricle; 2° ASD: atrial septal defect within the oval fossa

arterial abnormalities.[14–16] Even with such refinements, nonetheless, aortography is unlikely to yield information of the same diagnostic quality as selective coronary arteriography. In our group, paediatric cardiologists were catheterizing significantly younger patients than their adult cardiologic colleagues ($p < 0.0001$). It may be, therefore, that age played a part in the decision whether or not to perform selective coronary arteriography in the patients undergoing investigation. The finding, nonetheless, that the majority of abnormalities were congenital or surgically acquired, rather than being atherosclerotic, argues strongly for adequate imaging of the coronary arteries in all such patients.

The overall incidence of abnormalities identified in our patients was 27%, which is in accord with previous studies.[2,5] Certain malformations are more frequently associated with coronary arterial abnormalities. Patients with tetralogy of Fallot, for example, have been shown in previous angiographic studies to have an incidence of abnormal coronary arteries varying from 5 to 10%[17,18] to 36%.[19] In our study, one-fifth of patients with tetralogy of Fallot were found to have abnormal coronary arteries, with anomalous origin being the commonest finding. In two patients, a coronary artery had been occluded iatrogenically. Abnormalities were found in 7 of 40 patients with concordant atrioventricular and discordant ventriculo-arterial connections, and in 6 of 23 patients with discordant connections at both the atrioventricular and ventriculo-arterial junctions. These data, again, are in accord with previous studies.[20] In 58 patients with tetralogy of Fallot and complex pulmonary atresia, we found abnormalities in 7 (12%), these being acquired collateral arteries in four, and congenital anomalies in the other three.

Angiograms were reported as showing hypoplasia of the left anterior descending coronary artery in five patients. This is a lesion not well described in the literature. These arteries originated in a normal fashion and followed a normal course down the anterior interventricular groove, but were small and failed to reach the apex of the heart. Although three of the patients had had previous surgery, these arteries did not come to an abrupt halt but instead tapered in a normal fashion, and showed a normal pattern of branching. It did not look as though they had suffered iatrogenic occlusion at the time of surgery. In none of these patients was there any other apparent coronary arterial supply to the apex of the left ventricle. None of these patients suffered symptoms suggestive of myocardial ischaemia, though we have no objective data, for example from isotopic myocardial perfusion imaging, on the adequacy of perfusion of the left ventricular apex. It is noteworthy that no patient in our series had acquired atherosclerotic disease in the setting of cyanosis.

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In summary, therefore, just over one-quarter of our patients had abnormalities of their coronary arteries. Such abnormalities were found in one-third of patients undergoing selective coronary angiography without symptoms suggestive of coronary arterial disease. Haemodynamically significant lesions need to be identified at an early stage in order to treat symptoms and avoid progressive myocardial dysfunction, while the recognition of minor anomalies prior to surgical intervention may reduce the risk of iatrogenic injury. Injection in the aortic root is inadequate for visualization of the distal coronary arteries, whereas selective coronary angiography identifies asymptomatic coronary abnormalities in a high proportion of patients. Further studies are required to determine the risks and costs of selective coronary arteriography, and the potential benefits in terms of predicting morbidity and avoiding iatrogenic damage during cardiac surgery. An assessment could then be made of whether selective coronary arteriography should be recommended as a routine procedure during invasive investigation of grown-up patients with congenital cardiac disease.

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