Sudden cardiac death after repair of anomalous origin of left coronary artery from right sinus of Valsalva with an interarterial course

Case report and review of the literature

A. L. Nguyen · F. Haas · J. Evens ·
J. M. P. J. Breur

Published online: 9 October 2012
© Springer Media / Bohn Stafleu van Loghum 2012

Abstract Anomalous aortic origin of the coronary artery from the opposite sinus with interarterial course (AAOCA) is a rare condition with a high risk of sudden cardiac death (SCD) during or after strenuous exertion. SCD after repair of this anomaly is extremely rare. Here we present a 15-year-old athlete who collapsed on the basketball court in whom an anomalous origin of the left coronary artery from the right sinus of Valsalva with interarterial course (ALCA) was diagnosed. In spite of extensive pre-sport participation testing, SCD occurred shortly after surgical correction. We reviewed the literature to establish an evidence-based recommendation to aid physicians in conducting the optimal pre-sport participation management for the prevention of SCD in patients with a surgically corrected AAOCA/ALCA, especially for those who participate in strenuous exercise. Review of the literature (60 articles with 325 patients) reveals that post-surgical, pre-sport participation testing varies greatly but that mortality after surgical repair is extremely low (1.5 %). In conclusion, SCD can still rarely occur after repair of AAOCA despite extensive pre-sport participation testing. This should raise awareness among physicians treating these patients and

raises the question whether or not return-to-play guidelines need to be revised.

Keywords Anomalous coronary artery · Interarterial course · Sudden cardiac death · Surgery · Pre-sport participation testing

Introduction

An anomalous aortic origin of the coronary artery (AAOCA) is rare with a reported incidence of 0.3–1.3 % [1]. Anomalous left coronary artery from the right sinus with an interarterial course (ALCA) has a prevalence of 0.17 % and an annual risk at death of 0.24/100,000 per person-years [2]. Nevertheless anomalous coronary arteries are the second leading cause of sudden cardiac death (SCD) in Europe and North America with an incidence ranging from 12.2–17.2 % [3]. ALCA is most frequently associated with SCD during or after strenuous exercise [4]. SCD in these patients is likely to be the result of reduced diastolic coronary flow which may be explained by several mechanisms, namely interarterial compression during exertion, narrowing of the slit-like ostium, stenosis of the intramural course, acute angle take-off with possible torsion, spasm or kinking of the anomalous coronary artery [4, 5]. Considering the high risk of SCD, patients with an ALCA should undergo surgical correction [6]. There are several surgical techniques possible that tackle these underlying pathophysiological mechanisms such as fenestration or unroofing of the intramural component, re-implantation of the anomaly in the correct sinus, coronary artery bypass

A. L. Nguyen · J. M. P. J. Breur (✉)
Department of Paediatric Cardiology, Wilhelmina Children's
Hospital, University Medical Centre Utrecht,
PO Box 85090, 3508 AB Utrecht, the Netherlands
e-mail: h.breur@umcutrecht.nl

F. Haas · J. Evens
Department of Paediatric Cardiothoracic Surgery, Wilhelmina
Children's Hospital, University Medical Centre Utrecht,
PO Box 85090, 3508 AB Utrecht, the Netherlands

grafting and translocation of the pulmonary artery to prevent interarterial compression [5, 7–10]. From detection of AAOCA until surgical correction it is quite straightforward how to proceed. However, the diagnostics to be performed before a patient can return to play vary greatly in the literature.

Case report

A 15-year-old male patient, who played basketball at a high level, collapsed during a basketball match. He experienced chest pain, dizziness, nausea, vomiting and lost consciousness upon arrival of the ambulance. In retrospect he had experienced exertion-related chest pain once before. The patient had no coronary risk factors and there was no drug, doping or anabolic abuse. The patient's history revealed an immune complex glomerulonephritis at eight months with residual normal to high blood pressures and possible white coat hypertension at an age of two. Family history was negative for SCD or other familial diseases. At the referring hospital, the ECG showed a myocardial infarction with acute transmural anterolateral ischaemia and echocardiography revealed mild concentric left ventricle hypertrophy, most pronounced posteriorly, with normal systolic and diastolic function. Troponin reached a maximum of 6.23 µg/l (reference <0.10 µg/l). Conventional coronary angiography was inconclusive about a possible abnormal origination of the left main coronary artery (LMCA). Medical treatment was started and consisted of aspirin, plavix, simvastatin, metoprolol and fragmin and the patient was transferred to our paediatric heart centre. Cardiac magnetic resonance imaging/magnetic resonance angiogram was inconclusive in showing the origination of the coronary arteries; there was no delayed enhancement with normal function of the heart (ejection fraction 61 %). A computed tomography coronary angiography (CTCA) revealed an interarterial course of the LMCA with a proximal stenosis of 60 % (Fig. 1).

During surgery it became evident that the coronary artery had a slit-like orifice with intramural course (at least 2 mm) and interarterial course (at least 10 mm) before reaching the posterior aspect of the pulmonary artery.

The patient fully recovered with rapid normalisation of the troponin levels. Because of the high risk of recurrent ischaemia and SCD, an activity restriction was imposed upon surgical correction of the anomaly. Surgery consisted of an ostial plasty of the LMCA to widen the proximal 7 mm of the coronary artery. Additionally, a lateral pulmonary translocation was performed to decompress the interarterial segment [11]. Because of a narrow relation between the coronary ostia and the commissure, unroofing or coronary re-implantation were considered inadequate techniques as corrective procedures. Metoprolol and aspirin were prescribed

for 3 months and repeated echocardiography of the heart at 1 and 6 weeks postoperatively revealed good left ventricular function with a continuous orifice of the left coronary artery. One-month post surgery the patient underwent a stress single photon emission computed tomography (SPECT) myocardial perfusion with 52 mg adenosine infusion and a maximum of 80-Watt ergometric load for 6 minutes (max HR 139 bpm, max RR 140/70 mmHg), which showed no evidence of ischaemia. One month later the patient also underwent an exercise bicycle ergometry test because of persisting abnormal repolarisation with negative T waves in the left precordial leads. This test was performed according to the Godfrey protocol with 20 Watt/min at maximal exertion of 240 Watt (max HR 191 bpm, RERmax 1.17). The patient stopped because of leg pain. The test showed a normal exercise performance with normal resting blood pressure but an abnormal increase in blood pressure (RR syst >p95 at max exertion). Furthermore just before maximal exertion one premature ventricular contraction with left bundle branch block configuration was observed. There were no signs of ischaemia or arrhythmias. At this time our patient was symptom free and doing well. With these results the activity restriction was lifted. It was agreed to obtain BNP and troponin I the day after the exercise test. Unfortunately that same day, the patient collapsed during his first basketball training. Bystanders performed resuscitation and cardioversion was performed with an automated external defibrillator. On arrival to the emergency department the patient was breathing spontaneously and reactive with a weak pulse but he developed ventricular fibrillation shortly thereafter and died in spite of extensive therapy. Permission for post-mortem examination was not obtained.

Methods

A review of the literature concerning surgical repair of left main coronary artery anomalies from the right sinus of Valsalva with an interarterial course was performed by two independent reviewers (ALN, JMPJB). They compared results of this review and in case of differences in results they discussed inclusion or exclusion. Searches were performed in PubMed, Embase and the Cochrane database. The PubMed search consisted of three different searches: MESH terms “coronary vessel anomalies” AND “follow up” (664 hits), “coronary artery” AND “interarterial course” (73 hits) and (“coronary vessel anomalies”[Mesh] OR (“coronary artery” AND (anomalous[Title/Abstract] OR anomaly[Title/Abstract] OR anomalies[Title/Abstract])) OR “sinus of Valsalva/abnormalities”[Mesh]) AND (“follow up”[Mesh] OR “outcome”[Title/Abstract] OR “surgical repair”[Title/Abstract] OR “coronary vessel anomalies/complications”[Mesh] OR “coronary vessel anomalies/mortality”[Mesh] OR “coronary vessel anomalies/surgery”[Mesh]) NOT (“single common ostium”[Title/Abstract] OR “single coronary artery”[Title/

Fig. 1 CTCA of left main coronary artery originating from right sinus of Valsalva in a separate ostium near the right coronary artery. PA pulmonary artery; Ao Aorta; arrow interarterial course of left main coronary artery with proximal stenosis

Abstract] OR “pulmonary artery”[Title/Abstract] OR “stenting”[Title/Abstract] OR “stents”[Title/Abstract] OR “percutaneous”[Title/Abstract] OR “noncoronary”[Title/Abstract] OR “fistula”[Title/Abstract] OR “aneurysm”[Title/Abstract] OR “myocardial bridging”[Title/Abstract] OR “vascular fistula”[Mesh] OR “stents”[Mesh] OR “pulmonary artery”[Mesh] OR “coronary aneurysm”[Mesh] OR “myocardial bridging”[Mesh]) (1418 hits). All titles / abstracts were screened and 45 relevant articles were selected. This computer search was augmented by manual searches of reference lists from case reports, review articles and a systematic overview. Articles that concerned only right AAOCA, single coronary ostium and articles that did not report either postoperative diagnostics or mortality were excluded. This resulted in a total of 60 articles reporting on follow-up after correction of anomalous left main coronary artery from right sinus with interarterial course.

Results

Review of the literature

Review of the literature yielded 60 relevant articles with a total of 325 patients. The results are summarised in Table 1. Most patients underwent post-surgical testing. Tests that are most frequently used were echocardiography (59.1 %), stress ECG (27.4 %) and exercise or stress nuclear testing (17.2 %). Follow-up was often only briefly described. Rarely patients were restricted from activity after repair of the AAOCA. Only 2.2 % of the patients are symptomatic at the last follow-up and both morbidity and SCD are very low (9.2 % and 0.6 % (2 out of 325) respectively).

Discussion

Return-to-play recommendations

Guidelines have been established in order to prevent SCD among athletes with cardiovascular diseases. The 36th Bethesda Conference guideline is considered the gold standard in return-to-play decisions [68, 69]. According to this guideline patients with AAOCA should be excluded from participation in competitive sports because of increased risk of SCD during or after strenuous exertion. Patients are allowed to participate in all sports after surgical correction when two conditions are met: firstly when 3 months have passed after successful surgical correction of the anomaly and secondly when testing reveals no exercise-induced ischaemia, ventricular tachyarrhythmia or ventricular dysfunction during maximal exertion [69].

Patients who have suffered from myocardial infarction or underwent revascularisation are allowed to participate in athletic activity when their scars have healed and can tolerate strenuous activity (after 4 weeks). At that time the patient is at substantially increased risk of SCD when one of the following risk factors are present: impaired ejection fraction (<50 %), exercise-induced ischaemia or arrhythmias (including PCVs >10 % of beats/min, couplets or VT), or a significant degree of haemodynamic stenosis (>50 % of lumen). Patients with risk factors may therefore only participate in low static and low dynamic (class IA) competitive sports [69].

Patients with premature ventricular contractions without structural cardiac abnormalities may participate in all competitive sports, except when premature ventricular contractions increase upon exertion or when the patients experience exercise-induced symptoms of loss of consciousness, tiredness or dyspnoea [69].

Table 1 Overview of articles concerning anomalous aortic origin of coronary artery from opposite sinus of Valsalva with interarterial course

Authors	Type of article	No. of patients	No. LMCA from right sinus	No. with intra-mural course	No. of patients with symptoms	Postsurgical testing				Morbidity				FU in months (range)	LTFU	Symptomatic at last FU
						Echo	Stress echo	ECG	Stress ECG	CA	Cath	CTCA	Stress nuclear			
Mainwaring[12]	R	50	17	35	26	–	–	–	–	–	–	–	3	Median 5.7 yrs Mean 13.2 (max 14 yrs)	2	None
Davies[13]	R	36	15	34	29	–	–	–	–	1	–	–	–	6 ^b	None	1
Frommelt[14]	R	27	7	27	15	27	–	–	19	–	–	–	–	Mean 21 (1 wk–8.2 yrs)	None	None
Kaushal[15]	R	27	2	27	18	27	–	–	2	–	2	27	–	Mean 14	None	2
Brothers[16]	P	24	8	NM	17	24	24	–	24	–	–	–	24	Median 15 (2–48)	NM	None
Levin[17]	R	23	16	21	23	23	–	–	–	–	–	–	–	Mean 5.6 days (4–8)	None	NM
Muntaz[18]	R	22	7	22	21	22	4	–	1	–	1	6	6	Median 17 (1–63)	None	1
Gulati[10]	R	18	8	11	10	18	–	18	4	2	–	–	–	Mean 26.4 (1 wk–5 yrs)	NM	1
Romp[5]	P	9	6	9	8	9	8	–	9	–	2	–	1	Mean 29.3 (4–85)	None	None
elZem[19]	R	8	2	8	4	6	–	–	–	–	–	–	–	Mean 14 (1–39)	2	None
Erez[20]	R	8	4	3	8	8	–	8	–	–	–	–	8	Mean 12 (4–30)	None	None
Frommelt[21]	R	8	5	7	4	7	–	–	6	–	3	–	–	18	1	None
Osaki[22]	R	7	7	4	6	7	3	7	5	–	1	–	4	Median 23 (14–36)	None	1
Alphonso[11]	CR	4	2	3	4	–	1	–	3	1	1	–	–	18 (13–48)	None	None
Davis[23]	P	3	2	NM	2	2	–	–	3	–	–	–	1	12	None	NM
Donaldson [24]	R	3	3	3	3	–	–	–	3	3	–	–	–	10–14	None	None
Moodie[25]	CR	3	3	NM	3	1	–	–	1	–	1	–	1	Median 6 (2–10)	None	None
Mustafaj[26]	CR	2	2	1	2	–	–	1	2	2	2	–	–	Mean 30 (24–36)	None	None
Libertson[27]	R	2	2	NM	2	–	–	–	–	–	–	–	1	1 and 3 yrs	None	None
Nimdet[28]	CR	1	1	None	1	–	–	1	1	–	–	1	1	43	None	None
Ito[29]	CR	1	1	1	1	–	–	–	1	–	–	1	–	12	None	None
Pontone[30]	CR	1	1	NM	1	NM	–	–	–	–	–	–	–	NM	NM	NM
Brothers[31]	CR	1	1	1	1	1	–	–	–	–	–	–	1	3	None	None
Hosseini[32]	CR	1	1	1	1	1	–	–	–	–	–	–	–	12	None	None
Pellissier[33]	CR	1	1	NM	1	NM	–	–	–	–	–	–	–	12	None	NM
Guy [34]	CR	1	1	NM	1	NM	–	–	–	–	–	–	–	12	None	None
Lee[35]	CR	1	1	1	1	–	–	–	–	–	–	1	–	29	None	None
Slim[36]	CR	1	1	NM	1	–	–	–	–	1	–	–	1	NM	None	1
Absi[37]	CR	1	1	1	1	NM	–	–	–	–	–	–	–	4 days	None	NM
Bria[38]	CR	1	1	1	1	1	–	–	–	–	–	–	–	24	None	None
Emery[39]	CR	1	1	None	1	NM	–	–	–	–	–	–	–	NM	None	None
Gambetta[40]	CR	1	1	1	1	1	–	–	–	–	–	–	–	NM	None	None
Kupper[41]	CR	1	1	1	1	–	–	–	–	1	–	–	–	60	None	None
Vianna[42]	CR	1	1	NM	1	–	–	–	1	–	–	–	–	12	None	None
Angelini[43]	CR	1	1	1	1	–	–	–	–	–	–	–	1	12	None	None
Lopez[44]	CR	1	1	None	None	1	–	–	–	–	–	1	–	16	None	None

Table 1 (continued)

Authors	Type of article	No. of patients	No. LMCA from right sinus	No. with intra-mural course	No. of patients with symptoms	Postsurgical testing				Stress ECG	CA	Cath	CTCA	Stress nuclear	Morbidity	Mortality	FU in months (range)	LTFU	Symptomatic at last FU
						Echo	Stress echo	ECG	ECG										
Ou[45]	CR	1	1	NM	1	NM	–	1	1	–	–	–	–	–	None	None	NM	NM	NM
Kannan[46]	CR	1	1	1	1	1	–	–	–	–	–	–	–	–	None	None	5 days	None	NM
Sadaba[47]	CR	1	1	NM	1	1	–	–	–	–	–	–	–	–	None	None	12	None	None
Karamichalis[7]	CR	1	1	1	1	–	1	–	–	–	–	–	–	–	None	None	5	None	None
Khouzam[48]	CR	1	1	NM	1	NM	–	–	–	–	–	–	–	–	None	None	6	None	None
Nakajima[49]	CR	1	1	1	1	?	?	?	?	1	?	?	?	?	None	None	44	None	None
Patel[50]	CR	1	1	1	1	NM	–	–	–	–	–	–	–	–	None	None	NM	None	NM
Jureidini[51]	CR	1	1	1	1	1	–	–	–	1	1	–	–	1	None	None	5	None	NM
Phoon[52]	CR	1	1	1	1	1	–	–	–	–	–	–	–	–	1	None	NM	None	NM
Wang[53]	CR	1	1	1	1	NM	–	–	–	–	–	–	–	–	None	None	NM	None	None
Van Son[54]	CR	1	1	1	1	1	–	1	–	–	–	–	–	–	None	None	NM	None	NM
Manning[55]	CR	1	1	NM	1	–	–	1	–	–	–	–	–	1	None	None	6	None	None
Buenez[56]	CR	1	1	1	1	–	–	1	1	1	–	–	–	–	None	None	12	None	None
Selig[57]	CR	1	1	NM	1	NM	–	–	–	–	–	–	–	–	None	None	18	None	None
Alam[58]	CR	1	1	NM	1	NM	–	–	–	–	–	–	–	–	None	None	6	None	None
Amarasekera[59]	CR	1	1	1	1	1	–	–	–	1	–	–	–	–	None	None	12	None	None
Daliento[60]	CR	1	1	1	1	1	–	–	–	1	–	–	–	–	None	None	6	None	None
Salloun[61]	CR	1	1	NM	1	After stimulation no sign of myocardial ischaemia										NM	NM	None	NM
Thomas[62]	CR	1	1	NM	1	–	–	–	–	–	1 ^f	–	–	–	None	None	NM	None	None
Cohen[63]	CR	1	1	NM	1	–	–	–	–	–	1	–	–	1	1	None	9–14	None	None
Davia[64]	CR	1	1	NM	1	–	1	–	1	–	–	–	–	–	1	None	9 yrs	None	None
Grey[65]	CR	1	1	None	1	NM	–	–	–	–	–	–	–	–	None	None	8 days	None	NM
Sacks[66]	CR	1	1	1	1	–	–	–	1	–	1	–	–	–	None	None	5	None	None
Cheitlin[67]	CR	1	1	NM	1	NM	–	–	–	–	–	–	–	–	None	None	NM	None	None
Total (%)	3 P 13 R 44 CR	325 (100)	159 (48.9)	236 (72.6)	245 (75.4)	192 (59.1)	42 (12.9)	39 (12.0)	89 (27.4)	18 (5.5)	15 (4.6)	38 (11.7)	56 (17.2)	30 (9.2)	3 (0.9)	3 (0.9)	5 (1.5)	7 (2.2)	7 (2.2)

Cath cardiac catheterisation; *CA* coronary angiography; *CR* case report; *CTCA* computed tomography coronary artery; *ECG* electrocardiography; *Echo* echocardiography; *FU* follow-up; *LFTU* lost to follow-up; *NM* not mentioned; *P* prospective study; *R* retrospective review; *stress nuclear* nuclear (exercise or chemical) stress test; [?] additional testing mentioned without specifications; – not performed.

^a Including 1 heart transplantation 1 year after repair due to multiple prior infarctions.

^b Two patients required additional surgical interventions: catheter ablation for atrial fibrillation and aortic valve replacement due to aortic valve endocarditis.

^c Two ALCA patients required an additional surgical procedure; one was the first night postoperative for an aortic incision disruption and the other was two weeks postoperative due to localized pericardial effusion.

^d Including 1 heart transplantation, since the patient had an ejection fraction of 11–16 % and required a left ventricular assist device due to multiple infarctions.

^e A patient with severe aortic insufficiency was treated with aortic valve replacement with Ross procedure 44 months postoperatively.

^f This patient also underwent a myocardial metabolic study performed at rest and during atrial pacing to indicate ischaemia in relation to the coronary abnormality.

Applying return-to-play guidelines to case

Despite successful surgical correction and extensive diagnostic testing before returning to play our patient died from SCD during exertion. Not only does this raise questions regarding the cause of death, but this also questions whether or not the current guidelines sufficiently prevent SCD.

Our patient underwent extensive pre-sport participation testing. An echocardiogram was unremarkable. A combined exercise and chemically stressed myocardial perfusion SPECT showed no signs of ischaemia. A limitation of this test is the fact that the SPECT was performed after submaximal exercise with a maximal heart rate of 139 beats/min. However, our patient performed maximally during the exercise stress test (240 Watt, max HR 191, RERpeak 1.17). During or after maximal exercise no symptoms or signs of ischaemia (no pathological Q waves or ST segment abnormalities, no arrhythmias) were observed. According to the Bethesda guidelines our patient was allowed to participate in competitive sports again. The only criterion he did not fit was the 3-month postoperative interval. The guidelines do not motivate this time span and we strongly feel that after extensive pre-sports participation testing this patient was eligible to return to play two months postoperatively. Also risk stratification revealed that our patient was at low risk for SCD allowing him to participate in all competitive sports.

Since permission for post-mortem examination was not obtained, the cause of death remains uncertain in this patient. Sudden death may be explained in this patient either by a mechanical obstruction with secondary ischaemia and arrhythmia or by a primary arrhythmia. Obstruction could be caused by a thrombus; however, this patient was on aspirin and had no history of clotting disorders. Furthermore mechanical obstruction could be the result of progressive scarring postoperatively. This seems very unlikely, since our patient underwent an unremarkable exercise stress test on the day of his death. Finally, suboptimal surgery could result in residual obstructions; however, in this case we would expect evidence of ischaemia at echo, exercise stress test or stress myocardial perfusion SPECT. No arrhythmias were observed in the immediate postoperative period or during SPECT and exercise stress testing. The single premature ventricular contraction with left bundle branch block configuration during the exercise stress was considered a non-pathological premature ventricular contraction originating from the right ventricular outflow tract, an area of the heart that had not been ischaemic during the myocardial infarction. It is unlikely that at this time an arrhythmia originating from the right ventricular outflow tract caused SCD. On the other hand our patient did have mild concentric left ventricle hypertrophy associated with increased local oxygen consumption. This puts our patient at even more risk of ischaemia pre-operatively. After corrective surgery, perfusion of the thickened myocardium should have improved

as shown by the normal stressed myocardial perfusion SPECT and exercise stress test. Another possibility is that our patient developed an ischaemic scar during either his initial collapse or during the pre-sports participation testing. His baseline ECG never returned to normal. A final explanation could be that during basketball, associated with extreme peak performance, there was either ischaemia or a dynamic obstruction that could not be provoked by stress myocardial perfusion SPECT or exercise stress testing since these tests are endurance based.

Review of the literature shows that ALCA is rare and that 75 % of reported patients are symptomatic. Postoperative testing varies greatly in the literature and it is unclear (usually because it is not mentioned) whether or not the 36th Bethesda Conference recommendations are followed before returning to play. Furthermore, activity restrictions are hardly ever imposed postoperatively. Overall, follow-up was short with a wide range (4 days–14 years). SCD is extremely rare after repair of ALCA/AAOCA even in spite of the observation that in most cases exercise-induced ischaemia was not excluded optimally before returning to play. Davies and colleagues reported one patient who suffered from a postoperative subdural bleed after unroofing and a redo aortic valve replacement, who consequently died two months after at a rehabilitation centre [12]. Osaki and colleagues mentioned two deaths: one cardiac-related death concerns a 6-year-old girl who suddenly collapsed at home. She required resuscitation with extracorporeal membrane oxygenation and underwent emergency coronary re-implantation with patch enlargement. One day postoperatively she had poor ventricular function and a severe neurological insult after which care was withdrawn. The noncardiac death concerns a newborn with multiple congenital abnormalities who received palliative care [21]. Besides these two cardiac-related deaths that were reported among 325 successfully corrected patients, a survey performed by Brothers and colleagues mentioned two additional deaths after surgery. This survey was taken among 113 members of the Congenital Heart Surgeons' Society concerning treatment and management strategies for patients with AAOCA. A 7-year-old with ALCA died several weeks after an unroofing procedure and a 5-year-old died in the first postoperative week after undergoing an unknown type of surgical procedure. Unfortunately, limited information is available regarding the cause and circumstances leading to the deaths of these patients [70]. To our knowledge, this makes our patient the first patient ever reported with SCD during exercise after repair of ALCA/AAOCA.

Sensitivity postoperative testing

The 36th Bethesda Conference guidelines do not advise on the type of pre-sports participation testing. Exercise stress testing has a reported specificity of 50–74 % in detecting

coronary artery disease in patients with chest pain [71]. A stress exercise and dobutamine echocardiography have reported specificities of 82 % and 83 % respectively in detecting coronary artery disease [72]. SPECT and positron emission tomography (PET) have a reported specificity of 72 % and 90 % respectively in detecting coronary artery disease [73]. On the other hand a normal SPECT has a negative predictive value to rule out myocardial infarction of 98.8 % [74]. This implies that ischaemia can almost be ruled out with SPECT [75].

This contrasts with research performed by Brothers et al. who evaluated 24 corrected AAOCA patients with exercise stress test, stress echocardiography and myocardial perfusion scans and found evidence suggestive of ischaemia in nine postoperative patients with at least one of three testing modalities [15]. Six patients had normal myocardial perfusion scans and all patients were asymptomatic. This does not only show that patient complaints are not useful in detecting ischaemia but also that ischaemia may easily be missed when using only limited testing modalities. Therefore serial exercise stress tests, stress echocardiography and stress myocardial perfusion SPECT are recommended in order to adequately evaluate on-going ischaemia after AAOCA repair. Newer imaging technologies such as PET have proven higher sensitivity, specificity and accuracy in detection of coronary artery disease than SPECT and PET/CT [76]. A recent meta-analysis that studied the diagnostic accuracy of myocardial perfusion SPECT, PET and cardiac magnetic resonance confirms that PET has the highest diagnostic performance for detection of obstructive coronary artery disease; however, it also reveals cardiac magnetic resonance to be a good alternative since this has a similar diagnostic accuracy to PET and there is the advantage of no ionising radiation [77]. Unfortunately, to date there are no data on the accuracy of PET in detection of ischaemia after surgical repair of AAOCA [4]. Since optimal tests to evaluate surgical results are lacking and SCD does occur after correction of AAOCA, it seems mandatory to perform extensive and serial testing as described by Brothers et al. [15].

Lim and colleagues advocate that invasive testing with intravascular ultrasonography with dobutamine-stress testing and measurement of the fractional flow reserve is the most comprehensive test to assess obstructed coronary flow. They recommend this type of pre-surgical functional assessment for patients with AAOCA who are symptomatic or have positive non-invasive tests [4]. In the guidelines of ACC/AHA regarding management of congenital heart disease intravascular ultrasonography is also recommended for determining coronary flow restriction in patients with AAOCA (evidence level C) [6]. Considering the usefulness of these tests in the pre-surgical setting, one might advocate also applying this post-surgically and pre-sport. However, intravascular ultrasonography is an invasive technique with

practical limitations in smaller children and therefore this does not seem to be applicable in the paediatric population.

Recommendations

Patients with surgical repair of AAOCA should be offered serial testing with the most specific tests to rule out ischaemia before returning to play, especially when the patient performs strenuous exercise. In the current era this means repeated exercise stress tests, (stress) echocardiography and myocardial perfusion SPECT, PET/CT with maximal exercise or a stress cardiac magnetic resonance. In spite of extensive return-to-play testing SCD may occur in selected patients. Physicians treating these patients should be aware of that.

Conclusion

Extensive return-to-play testing cannot prevent SCD in selected patients after uneventful surgical repair of AAOCA. This should raise awareness among physicians treating these patients and raises the question whether or not these return-to-play guidelines need to be revised.

Competing interests None.

Funding None.

References

1. Ugalde H, Ramirez A, Ugalde D, et al. Coronary artery origin anomalies. Analysis of 10,000 coronary angiographies. *Rev Med Chill*. 2010;138:7–14.
2. Friedman AH, Silverman NH. Congenital anomalies of the coronary arteries. In: Anderson RH, editor. *Paediatric cardiology*. Philadelphia: Churchill Livingstone; 2009. p. 933–43.
3. Arrigan MT, Killeen RP, Dodd JD, et al. Imaging spectrum of sudden athlete cardiac death. *Clin Radiol*. 2011;66:203–23.
4. Lim JCE, Beale A, Ramcharitar S. Anomalous origination of a coronary artery from the opposite sinus. *Nat Rev Cardiol*. 2011;8:706–19.
5. Romp RL, Herlong JR, Landolfo CK, et al. Outcome of unroofing procedure for repair of anomalous origin of left or right coronary artery. *Ann Thorac Surg*. 2003;76:589–95.
6. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation*. 2008;118:714–833.
7. Karamichalis JM, Vircella LA, Murphy DJ, et al. Simplified technique for correction of anomalous origin of left coronary artery from the anterior aortic sinus. *Ann Thorac Surg*. 2003;76:266–7.
8. Krasuski RA, Magyar D, Hart S, et al. Long-term outcome and impact of surgery on adults with coronary arteries originating from the opposite coronary cusp. *Circulation*. 2011;123:154–62.

9. Rodefeld MD, Culbertson CB, Rosenfeld HM, et al. Pulmonary artery translocation: a surgical option for complex anomalous coronary artery anatomy. *Ann Thorac Surg.* 2007;72:2150–2.
10. Gulati R, Reddy VM, Culbertson C, et al. Surgical management of coronary artery arising from the wrong coronary sinus, using standard and novel approaches. *J Thorac Cardiovasc Surg.* 2007;134:1171–8.
11. Alphonso N, Anagnostopoulos PV, Nölke L, et al. Anomalous coronary artery from the wrong sinus of Valsalva: a physiologic repair strategy. *Ann Thorac Surg.* 2007;83:1472–6.
12. Mainwaring RD, Reddy VM, Reinhartz O, et al. Anomalous aortic origin of coronary artery: medium-term results after surgical repair in 50 patients. *Ann Thorac Surg.* 2011;92:691–7.
13. Davies JE, Burkhart HM, Dearani JA, et al. Surgical management of anomalous aortic origin of coronary artery. *Ann Thorac Surg.* 2009;88:844–8.
14. Frommelt PC, Sheridan DC, Berger S, et al. Ten-year experience with surgical unroofing of anomalous aortic origin of a coronary artery from the opposite sinus with an interarterial course. *J Thorac Cardiovasc Surg.* 2011;142:1046–51.
15. Kaushal S, Backer CL, Popescu AR, et al. Intramural coronary length correlates with symptoms in patients with anomalous aortic origin of the coronary artery. *Ann Thorac Surg.* 2011;92:986–91. discussion 991–2.
16. Brothers JA, McBride MG, Seliem MA, et al. Evaluation of myocardial ischaemia after surgical repair of anomalous aortic origin of coronary artery in a series of pediatric patients. *J Am Coll Cardiol.* 2007;50:2078–82.
17. Levin R, Degrange M, Lezana F, et al. Diagnosis and surgical treatment of the anomalous origin of the coronary arteries. *Revist Argent Cardiol.* 2011;79:125–31.
18. Mumtaz MA, Lorber RE, Arruda J, et al. Surgery for anomalous aortic origin of the coronary artery. *Ann Thorac Surg.* 2011;91:811–4. discussion 814–5.
19. elZein C, Hanhan Z, Massad M, et al. Surgical repair of anomalous coronary arteries arising from the opposite sinus of Valsalva in infants and children. *J Card Surg.* 2009;24:466–9.
20. Erez E, Tam VK, Dublin NA, et al. Anomalous coronary artery with aortic origin and course between the great arteries: improved diagnosis, anatomic findings, and surgical treatment. *Ann Thorac Surg.* 2006;82:973–7.
21. Frommelt PC, Frommelt MA, Tweddell JS, et al. Prospective echocardiographic diagnosis and surgical repair of anomalous origin of a coronary artery from the opposite sinus with an interarterial course. *J Am Coll Cardiol.* 2003;42:148–54.
22. Osaki M, McCrindle BW, Van Arsdell G, et al. Anomalous origin of coronary artery from the opposite sinus of Valsalva with an interarterial course: clinical profile and approach to management in de pediatric population. *Pediatr Cardiol.* 2008;29:24–30.
23. Davis JA, Cecchin F, Jones TK, et al. Major coronary artery anomalies in a pediatric population: incidence and clinical importance. *J Am Coll Cardiol.* 2001;37:593–7.
24. Donaldson RM, Raphael M, Radley-Smith R, et al. Angiographic identification of primary coronary anomalies causing impaired myocardial perfusion. *Cathet Cardiovasc Diagn.* 1983;9:237–49.
25. Moodie DS, Gill C, Loop FD, et al. Anomalous left main coronary artery originating from right sinus of Valsalva: pathophysiology, angiographic definition and surgical approaches. *J Thorac Cardiovasc Surg.* 1980;80:198–205.
26. Mustafa I, Gula G. Anomalous origin of the left coronary artery from anterior aortic sinus: a potential cause of sudden death. *J Thorac Cardiovasc Surg.* 1981;82:297–300.
27. Liberthson RR, Dinsmore RE, Fallon JT. Aberrant coronary artery origin from the aorta. Report of 18 patients, review of literature and delineation of natural history and management. *Circulation.* 1979;59:748–54.
28. Nimdet K, Soongswang J, Sriyodchati S: An interarterial course of anomalous coronary artery and an alternative surgical option. *Cardiol Young.* Published Online First: 9 February 2012. doi:10.1017/S1047951111002113.
29. Ito H, Takabayashi S, Kajimoto M, et al. Patch angioplasty and neo-ostium creation for intramural left coronary artery. *Interact Cardiovasc Thorac Surg.* 2011;13:661–3.
30. Pontone G, Andreini D, Bertella E, et al. Detection of left main coronary artery anomalous origin with low-dose multidetector computed tomography using prospective ECG gating. *J Cardiovasc Med (Hagerstown).* 2011;12:506–9.
31. Brothers J, Carter C, McBride M, Spray T, et al. Anomalous left coronary artery origin from the opposite sinus of Valsalva: evidence of intermittent ischaemia. *J Thorac Cardiovasc Surg.* 2010;140:27–9.
32. Hosseini MT, Kourliouros A, Sarsam M. Aberrant left coronary artery, modified unroofing technique, and patch angioplasty. *J Card Surg.* 2010;25:508–10.
33. Pellissier A, Le Pennec V, Labombarda F. Anomalous aortic origin of left coronary artery revealed by ventricular fibrillation. *Pediatr Cardiol.* 2010;31:556–7.
34. Guy TS, Tseng E, Ratcliffe MB, et al. A simple correction for anomalous coronary arteries in adults. *J Thorac Cardiovasc Surg.* 2009;137:50–1.
35. Lee MK, Choi JB, Kim KH, et al. Surgery for anomalous origin of the left main coronary artery from the right sinus of Valsalva in association with left main stenosis. *Tex Heart Inst J.* 2009;36:309–12.
36. Slim A, Thurlow J, Blevins J, et al.: Discovery of a symptomatic left anomalous coronary artery from the opposite sinus and post-operative considerations. *Case Report Med.* Published Online 15 October 2009. doi:10.1155/2009/509064.
37. Absi MA, Mathis C, Chase N. Anomalous aortic origin of the left coronary artery. *Cardiol Young.* 2008;18:543–5.
38. Bria S, Chessa M, Abella R, et al. Aborted sudden death in a young football player due to anomalous origin of the left coronary artery: successful surgical correction. *J Cardiovasc Med.* 2008;9:834–8.
39. Emery M, Ghumman W, Teague S, et al. A hazardous finding of a rare anomalous left main coronary artery in a patient with a secundum atrial septal defect. *Vasc Health Risk Manag.* 2008;4:259–62.
40. Gambetta K, Cui W, El-Zein C, et al. Anomalous left coronary artery from the right sinus of Valsalva and noncompaction of the left ventricle. *Pediatr Cardiol.* 2008;29:434–7.
41. Kupper MS, Bethuyn N, Rubay J, et al. Aberrant left coronary artery arising from the right sinus of Valsalva: case reports of a rare entity. *J Thorac Cardiovasc Surg.* 2008;136:788–9.
42. Vianna CB, Gonzalez MM, Dallan LA, et al. Anomalous coronary artery causing transmural ischaemia and ventricular tachycardia in a high school athlete. *Resuscitation.* 2007;74:183–6.
43. Angelini P, Walmsley RP, Liberos A, et al. Symptomatic anomalous origination of the left coronary artery from the opposite sinus of Valsalva. Clinical presentations, diagnosis, and surgical repair. *Tex Heart Inst J.* 2006;33:171–9.
44. Lopez L, Mercer-Rosa L, Zahn EM, et al. The “hinge-twist” technique for anomalous origin of the left coronary artery. *Ann Thorac Surg.* 2006;82:19–21.
45. Ou P, Iserin F, Vouhe P, et al. Anomalous origin of the left coronary artery from the right aortic sinus: surgery based on diagnosis by 64-slice CT. *Eur J Cardiothorac Surg.* 2006;29:610.
46. Kannam HC, Satou G, Gandelman G, et al. Anomalous origin of the left main coronary artery from the right sinus of Valsalva with an intramural course identified by transesophageal echocardiography in a 14 year old with acute myocardial infarction. *Cardiol Rev.* 2005;13:219–22.
47. Sadaba JR, Nair UR. Selective arterialization of the coronary venous system. *Ann Thorac Surg.* 2004;78:1458–60.

48. Khouzam R, Marshall T, Lowell D, et al. Left coronary artery originating from right sinus of Valsalva with diagnosis confirmed by CT—a case report. *Angiology*. 2003;54:499–502.
49. Nakajima H, Yagihara T, Uemura H, et al. Extended unroofing procedure for creation of a new ostium for anomalous left coronary artery. *Ann Thorac Surg*. 2001;72:1768–9.
50. Patel K, Davidson A, Karl TR. Anomalous left coronary artery arising from the right coronary cusp. *Ann Thorac Surg*. 2001;71:2045.
51. Jureidini SB, Singh GK, Marino CJ, et al. Aberrant origin of the left coronary artery from the right aortic sinus: surgical intervention based on echocardiographic diagnosis. *J Am Soc Echocardiogr*. 2000;13:1117–20.
52. Phoon CK, Van Son J, Moore PA, et al. Aberrant left coronary artery arising from the right sinus of Valsalva with a right coronary arteriovenous malformation. *Pediatr Cardiol*. 1997;18:385–8.
53. Wang A, Pulsipher MW, Jaggars J, et al. Simultaneous biplane coronary and pulmonary arteriography: a novel technique for defining the course of an anomalous left main coronary artery originating from the right sinus of Valsalva. *Cathet Cardiovasc Diagn*. 1997;42:73–8.
54. Van Son JA, Haas GS. Anomalous origin of left main coronary artery from right sinus of Valsalva: modified surgical treatment to avoid neo-coronary ostial stenosis. *Eur J Cardiothorac Surg*. 1996;10:467–9.
55. Manning WJ, Li W, Cohen SI, et al. Improved definition of anomalous left coronary artery by magnetic resonance coronary angiography. *Am Heart J*. 1995;130:615–7.
56. Bucsenec D, Messmer BJ, Gillor A, et al. Management of anomalous origin of the left coronary artery from the right sinus of Valsalva. *J Thorac Cardiovasc Surg*. 1994;107:1370–3.
57. Selig MB, Jafari N. Anomalous origin of the left main coronary artery from the right coronary artery ostium-interarterial subtype: angiographic definition and surgical treatment. *Cathet Cardiovasc Diagn*. 1994;31:41–7.
58. Alam M, Brymer J, Smith S. Transesophageal echocardiographic diagnosis of anomalous left coronary artery from the right aortic sinus. *Chest*. 1993;103:1617–8.
59. Amarasena NL, Pillai RP, Forfar JC. Atypical ventricular tachycardia and syncope with left coronary artery origin from the right coronary sinus. *Br Heart J*. 1993;70:391–2.
60. Daliento L, Fasoli G, Mazzucco A. Anomalous origin of the left coronary artery from the anterior aortic sinus: role of echocardiography. *Int J Cardiol*. 1993;38:89–91.
61. Salloum JA, Thomas D, Evans J. Transoesophageal echocardiography in diagnosis of aberrant coronary artery. *Int J Cardiol*. 1991;32:106–8.
62. Thomas D, Salloum J, Montalescot G, et al. Anomalous coronary arteries coursing between the aorta and pulmonary trunk: clinical indications for coronary artery bypass. *Eur Heart J*. 1991;12:832–4.
63. Cohen AJ, Grishkin BA, Helsel RA, et al. Surgical therapy in the management of coronary anomalies: emphasis on utility of internal mammary artery grafts. *Ann Thorac Surg*. 1989;47:630–7.
64. Davia JE, Green DC, Cheitlin MD, et al. Anomalous left coronary artery origin from the right coronary sinus. *Am Heart J*. 1984;108:165–6.
65. Grey DP, Koster Jr JK, Farrell PW, et al. Surgical correction of anomalous left coronary artery from the anterior sinus of Valsalva. *Tex Heart Inst J*. 1984;11:182–6.
66. Sacks JH, Londe SP, Rosenbluth A, et al. Left main coronary bypass for aberrant (aortic) intramural left coronary artery. *J Thorac Cardiovasc Surg*. 1977;73:733–7.
67. Cheitlin MD, De Castro CM, McAllister HA. Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva, a not-so-minor congenital anomaly. *Circulation*. 1974;50:780–7.
68. Oliveira LP, Lawless CE. Making prudent recommendations for return-to-play in adult athletes with cardiac conditions. *Curr Sports Med Rep*. 2011;10:65–77.
69. Maron BJ, Zipes DP. 36th Bethesda Conference: Eligibility recommendations for competitive athletes with cardiovascular abnormalities. *J Am Coll Cardiol*. 2005;45:1313–75.
70. Brothers J, Gaynor JW, Paridon S, et al. Anomalous aortic origin of a coronary artery with an interarterial course: understanding current management strategies in children and young adults. *Pediatr Cardiol*. 2009;30:911–21.
71. Gibbons RJ, Balady GJ, Bricker JT, et al. ACC/AHA 2002 guideline update for exercise testing: summary article a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee to Update the 1997 Exercise Testing Guidelines). *Circulation*. 2002;106:1883–92.
72. Pellikka PA. Stress echocardiography in the evaluation of chest pain and accuracy in the diagnosis of the coronary artery disease. *Prog Cardiovasc Dis*. 1997;39:523–32.
73. Vesely MR, Dilsizian V. Nuclear cardiac stress testing in the era of molecular medicine. *J Nucl Med*. 2008;49:399–413.
74. Metz LD, Beattie M, Hom R, et al. The prognostic value of normal exercise myocardial perfusion imaging and exercise echocardiography. A Meta-analysis. *J Am Coll Cardiol*. 2007;49:227–37.
75. Beller GA, Heede RC. SPECT imaging for detecting coronary artery disease and determining prognosis by noninvasive assessment of myocardial perfusion and myocardial viability. *J Cardiovasc Transl Res*. 2011;4:416–24.
76. Al Moudi M, Sun Z, Lenzo N. Diagnostic value of SPECT, PET and PET/CT in the diagnosis of coronary artery disease: a systematic review. *Biomed Imaging Interv J*. 2011;7:9.
77. Jaarsma C, Leiner T, Bekkers SC. Diagnostic performance of noninvasive myocardial perfusion imaging using single-photon emission computed tomography, cardiac magnetic resonance, and positron emission tomography imaging for the detection of obstructive coronary artery disease: a meta-analysis. *J Am Coll Cardiol*. 2012;59:1719–28.