Surgical repair of anomalous aortic origin of a coronary artery†

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Abstract

OBJECTIVES: Anomalous aortic origin of a coronary artery (AAOCA) is a rare congenital heart defect that has been associated with myocardial ischaemia and sudden death. There is an ongoing controversy over the indications for surgical intervention and the efficacy of that treatment compared with the natural history. The purpose of this study was to evaluate the medium-term results of surgical repair of AAOCA.

METHODS: Seventy-six patients underwent surgical repair of AAOCA at our institution from 1999 to 2013. There were 55 males and 21 females, and the median age at surgery was 15 years. Forty-seven (62%) of the 76 patients had an anomalous right coronary artery, 27 had an anomalous left coronary and 2 had an eccentric single coronary ostia. Forty-one patients had preoperative symptoms of myocardial ischaemia.

RESULTS: Surgical repair was accomplished by unroofing of an intramural coronary in 55, reimplantation in 7 and pulmonary artery translocation in 14. There has been no early or late mortality, with a median duration of follow-up of 6 years. One patient presented with severe myocardial ischaemia and subsequently underwent heart transplantation a year following AAOCA surgery. The remaining patients have all remained free of cardiac symptoms.

CONCLUSIONS: The results of this study demonstrate two major principles. First, surgical repair of AAOCA is quite safe in centres that take care of a significant number of patients with this entity. Secondly, the surgery is highly effective in eliminating symptoms of myocardial ischaemia. The growing amount of data on postoperative patients suggests that surgical repair can prevent the adverse events seen in the untreated ‘natural’ history. Based on these observations, it is our current recommendation that all teenagers identified with AAOCA should undergo surgical repair.

Keywords: Congenital heart defect (coronary arteries) • Coronary artery disease • Coronary artery imaging • Myocardial ischaemia

INTRODUCTION

Anomalous aortic origin of a coronary artery (AAOCA) from the wrong sinus of Valsalva is a relatively rare congenital heart defect (CHD). It was originally described in association with sudden death in young athletes [1, 2]. Over the ensuing years, the diagnosis of AAOCA was made with increasing frequency in patients presenting with symptoms of myocardial ischaemia including chest pain, syncope and heart failure [3–5]. In addition, it has become evident that AAOCA is associated with concomitant CHDs [6] and a familial incidence has also been described [7]. Based on differences in anatomic patterns, we have suggested that sudden death and the other modes of presentation may represent separate ‘diseases’ sharing a common underlying anatomy [8]. Although the diagnosis of AAOCA would imply that it is a single entity, it has become increasingly clear that there are multiple subgroups defined by the underlying anatomic patterns. The most obvious distinction is the site of origin (right vs left) of the anomalous coronary arteries. There is also increasing evidence to suggest that the presence or absence of a proximal intramural coronary course may have a strong influence on clinical presentation [8, 9]. These two parameters (right vs left, intramural vs non-intramural) create a two-by-two box diagram with four identifiable subgroups. These four subgroups have distinctly different modes of presentation and incidence, and it is likely that they may also have very different risk profiles with regard to their natural history.

There is currently a controversy over the indications for surgery in patients with AAOCA. The consensus statement from the
American College of Cardiology and American Heart Association would support that surgery is indicated in patients with an anomalous left coronary due to the risk of sudden death and in patients with an anomalous right coronary who present with symptoms of myocardial ischaemia [10]. Patients with concomitant CHD have an independent indication for surgery. Therefore, the controversy is narrowed to whether patients who have an anomalous right coronary and are asymptomatic should undergo surgical repair. Brothers et al. [11] have concluded that asymptomatic patients with an anomalous right coronary do not warrant surgery. However, a subset of asymptomatic patients with anomalous right coronary will have an intramural course. In a previously published study, our group showed that the presence of an intramural course was associated with symptoms in 87% of patients [8]. Based on these data, we have suggested that the presence of an intramural origin of a right coronary artery should be an additional indication for surgical repair. Thus, the indications for AAOCA remain controversial, with a consensus regarding the majority of patients but a lack of consensus pertaining to a small but important minority.

The purpose of this study was to review our surgical experience with AAOCA to evaluate the patterns of anatomy, presentation and medium-term outcomes. It is a specific goal to elucidate additional insights regarding the controversial aspects of surgery for AAOCA as to who should undergo this surgical repair.

**METHODS**

This study is a retrospective review of 76 patients who underwent surgical repair of AAOCA at Lucile Packard Children’s Hospital, Oakland Children’s Hospital and Children’s Hospital of Central California. The operations were performed by Stanford University faculty members, and the period encompasses January 1999 through June 2013. The study was approved by the Institutional Review Board at each of the respective institutions.

There were 55 males and 21 females included in this study. The median age at the time of surgery was 16 years. There were 17 patients between the ages of 0 and 10 years, 54 patients between the ages of 10 and 20 years 4 patients between the ages of 20 and 25 years, and one 47-year old patient.

There were four separate modes of presentation, as summarized in Fig. 1. Forty-one patients presented with symptoms of myocardial ischaemia. Eighteen patients had CHDs as their primary mode of presentation, and were subsequently found to have AAOCA. These CHDs are given in Table 1. Sixteen patients were asymptomatic, did not have CHDs and were discovered to have AAOCA during screening evaluations for murmurs, palpitations or dizziness. Finally, there was one set of siblings in whom the diagnosis of AAOCA was recognized in one child and then identified in his asymptomatic brother [12].

The symptomatic patients had a variety of different forms of presentation. The most common presenting symptom was chest pain with exertion, which occurred in 29 of these 41 patients. Syncope or presyncope was the presenting symptom in 12 patients, while 3 patients had symptoms of dyspnoea or heart failure. None of the patients included in this study was a survivor of a sudden death event who then received successful cardiopulmonary resuscitation.

Of the 41 symptomatic patients, there were 20 patients who had evidence of having sustained ischaemic myocardial damage prior to their diagnosis of AAOCA. This included 12 patients with an abnormal electrocardiogram (EKG), 4 patients who had documented leakage of cardiac enzymes and 7 patients who had functional abnormalities. The presence of functional abnormalities was documented by echocardiography in 7 of the 7 and computed tomography (CT) in 3 of 3. All 7 patients with functional abnormalities had symptoms of chest pain with exertion prior to their diagnosis, and 3 had progressed to dyspnoea or heart failure. Of note, 18 of the 20 patients with ischaemic myocardial damage came from the cohort of 29 patients with chest pain, while there were only 2 patients (out of 12) with syncope or presyncope who had evidence of myocardial damage.

A comparison of patients who presented with symptoms with those without symptoms reveals similarities and dissimilarities. The median age at surgery for the 41 symptomatic patients was 16 years (range 8–47 years) and the median age of surgery for the 16 asymptomatic patients was 17 years (range 12–22 years). Twenty of the 41 symptomatic patients (48%) had some evidence of ischaemic myocardial damage. In contrast, none of the asymptomatic patients had experienced myocardial damage.

The majority of patients have been followed by the cardiology groups at Lucile Packard Children’s Hospital, Oakland Children’s Hospital or Children’s Hospital of Central California. We have lost contact with 4 patients over the years; two families moved and were subsequently lost to follow-up and 2 patients graduated from paediatric care and entered into the domain of adult medical care. All 4 of these patients had been followed for a considerable period of time following their surgery, and had not manifested any symptoms or other notable problems. The duration of follow-up before

<table>
<thead>
<tr>
<th>CHDs and AAOCA identified preoperatively</th>
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<tbody>
<tr>
<td>Ventricular septal defect</td>
</tr>
<tr>
<td>Atrial septal defect</td>
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<tr>
<td>Coronary-cameral fistula</td>
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<tr>
<td>CHDs known, AAOCA identified intraoperatively</td>
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<tr>
<td>Transposition of the great arteries</td>
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<tr>
<td>Ventricular septal defect</td>
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<tr>
<td>Tetralogy of Fallot</td>
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<tr>
<td>Truncus arteriosus</td>
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<tr>
<td>Bicuspid aortic valve</td>
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<td>Subaortic membrane</td>
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Figure 1: Pie chart demonstrating the mode of presentation for the 76 patients who underwent surgical repair of AAOCA. Fifty-four percent of the cohort were symptomatic and 24% had AAOCA and associated CHDs. Twenty-one percent of patients were asymptomatic and did not have CHDs, and there was one set of siblings diagnosed with AAOCA.
contact was lost was 2–4 and 7 years following surgery, and for these 4 patients, the duration of follow-up, date of surgery to the date of last contact, was considered to be the point of last contact.

Statistical analysis was performed using the Test of Independent Proportions to compare binomial functions of the two study groups. A $P$-value of $<0.05$ was considered statistically significant.

RESULTS

There was no operative mortality in this cohort of 76 patients undergoing AAOCA surgery. None of the patients required reoperation for revision or bleeding, and none of the patients had evidence of having sustained perioperative myocardial injury. The median length of hospital stay was 5 days. There were 8 surgical complications, including development of pleural effusions in 3, post-cardiotomy syndrome in 4 and transient heart block in 1.

Coronary anatomy

Forty-seven of the 76 patients had an anomalous right coronary artery arising from the left sinus of Valsalva. Twenty-seven patients had an anomalous left coronary arising from the right sinus of Valsalva. Two patients had a single coronary ostium that was directly above the commissure between the right and left sinuses of Valsalva.

Fifty-five patients had an intramural proximal coronary course. Of these, 38 were anomalous right coronaries and 17 were anomalous left coronary arteries. Stated differently, 38 of 47 (81%) anomalous right coronaries were intramural, and 17 of 27 (63%) anomalous left coronaries were intramural. A total of 17 patients had a single coronary origin, of whom 3 had an intramural course. These data are summarized in Table 2.

Choice of operation

The 55 patients who had an intramural proximal coronary all underwent an unroofing procedure. Among the 21 patients who did not have an intramural proximal coronary, 7 had two separate coronary ostia, and these patients underwent coronary transfer with reimplantation into the proper sinus of Valsalva. There were 14 patients who did not have an intramural proximal coronary and had a single coronary ostium, and these patients underwent a pulmonary artery translocation procedure. This surgical algorithm is depicted in Fig. 2.

Correlation between anatomy and symptoms

There were a total of 41 patients who presented with preoperative symptoms of myocardial ischaemia, including chest pain, syncope or heart failure. Conversely, there were 16 patients who were asymptomatic and had the diagnosis of AAOCA made during the screening evaluation for murmurs or palpitations. Figure 3 demonstrates the $2 \times 2$ box diagram of these 57 patients and the correlation between anatomy and symptoms. Patients with an intramural proximal coronary had an 85% prevalence of symptoms, compared with a 41% prevalence in patients without an intramural course ($P < 0.05$).

Medium-term follow-up

There have been no late deaths in this cohort of 76 patients who underwent AAOCA surgery. The patients have been followed for

Table 2: Summary of the coronary artery patterns identified intraoperatively

<table>
<thead>
<tr>
<th>Coronary pattern</th>
<th>$n$</th>
<th>Percent</th>
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<tbody>
<tr>
<td>Interarterial</td>
<td>76</td>
<td>100%</td>
</tr>
<tr>
<td>Intramural</td>
<td>55</td>
<td>72%</td>
</tr>
<tr>
<td>Anomalous right coronary</td>
<td>47</td>
<td>62%</td>
</tr>
<tr>
<td>Single coronary ostium</td>
<td>8</td>
<td>11%</td>
</tr>
<tr>
<td>Anomalous left coronary</td>
<td>27</td>
<td>35%</td>
</tr>
<tr>
<td>Single coronary ostium</td>
<td>7</td>
<td>11%</td>
</tr>
<tr>
<td>Eccentric location above commissure</td>
<td>2</td>
<td>3%</td>
</tr>
<tr>
<td>Single coronary ostium</td>
<td>2</td>
<td>3%</td>
</tr>
<tr>
<td>Single coronary ostium</td>
<td>17</td>
<td>22%</td>
</tr>
<tr>
<td>Normal location, intramural</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Normal location, not intramural</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>Eccentrically located</td>
<td>2</td>
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an average of 6 years (range 1 month to 14 years) or a total of 424 patient-years. There have been no late sudden death events, and none of the patients has demonstrated any evidence of new myocardial ischaemia. One patient, who had sustained severe myocardial damage prior to the diagnosis of AAOCA, did not show any improvement in left ventricular function following surgery and required cardiac transplantation 1 year later. There were 2 patients in this series who experienced ‘pain in their chest’ ~1 year after surgery, and because of their underlying diagnosis, underwent an extensive evaluation (including EKG, echocardiogram, CT angiogram, nuclear medicine perfusion study and cardiac catheterization). In both patients, all of the studies were entirely normal and it was concluded that the aetiology of their chest pain was not related to myocardial ischaemia.

DISCUSSION

This study summarizes our experience with 76 patients who underwent surgical repair of AAOCA. There was no operative mortality, nor has there been any late mortality. None of the patients has experienced any late sudden death events. In addition, none of the postoperative patients has had cardiac symptoms attributable to myocardial ischaemia, including the 41 patients who had symptoms of cardiac ischaemia preoperatively. These data indicate two important principles: (i) surgical repair of AAOCA is quite safe in centres performing a significant volume of AAOCA surgery and (ii) surgical repair is highly efficacious in eliminating the symptoms of myocardial ischaemia. This cohort now has an excess of 400 patient-years of follow-up, and suggests that AAOCA surgery can prevent the adverse events seen in the untreated ‘natural’ history of this disease.

The diagnosis of AAOCA is quite diverse with respect to the anatomy of the coronary arteries. Important variables include the site of origin (right vs left), proximal anatomy (intramural vs non-intramural) and presence of two coronary ostia vs a single coronary ostium. In the current series, approximately two-thirds of the patients had an anomalous right coronary artery, which is in accordance with many other studies of this subject [13–16]. It is worth noting that anomalous right coronary arteries are about six times more common than anomalous left coronaries [17], so that there is a three-fold disparity between the anatomic incidence and the clinical incidence. We observed an intramural proximal coronary course in 72% of our patients, and conversely, 28% of the patients did not have an intramural course. Two coronary ostia were observed in 59 of 76 (78%) patients, while a single coronary ostium was identified in 17 (22%) patients. These findings highlight the many different anatomic forms of the diagnosis of AAOCA.

Our surgical algorithm is based on the intraoperative findings of the coronary anatomy and is summarized in Fig. 2. The 55 patients

![Figure 3](image-url) Figure 3: Two-by-two box diagram demonstrating the distribution of anatomy of patients with AAOCA. Half of the entire cohort of AAOCA patients had an intramural right coronary artery. In parentheses are the number of symptomatic patients divided by the number of symptomatic plus asymptomatic patients. We have excluded from this analysis the patients whose primary indication for surgery was associated CHDs and the one sibling diagnosed with AAOCA.

![Figure 4](image-url) Figure 4: (A) Illustration of the lateral PA translocation procedure. This procedure is ideally suited for an anomalous right coronary that originates from a common, single ostium and is not intramural. (B) The main PA is transected at the bifurcation and moved to the patient’s left.
who had a proximal intramural course all underwent an unroofing procedure (49 had a conventional unroofing and 6 had a ‘fenestration’ created due to the juxtaposition of the intramural segment with the commissure). Of the 21 patients who did not have an intramural segment, 7 had separate coronary ostia and therefore underwent a coronary transfer. Fourteen patients had a single coronary ostium without an intramural segment and underwent a pulmonary translocation procedure [18, 19]. There were an additional 4 patients who underwent a pulmonary artery translocation concomitantly with unroofing when the intramural segment was relatively short (<2 mm). In this situation, unroofing leaves a segment of the proximal coronary that remains interarterial, and we have supplemented the unroofing with a concomitant pulmonary translocation procedure.

We have described two different pulmonary artery translocation procedures for the circumstance of an interarterial coronary that is not intramural and does not have a separate coronary ostia. The lateral pulmonary artery translocation is performed by dividing the main pulmonary artery at its bifurcation to the right and left branch pulmonary arteries. The main pulmonary artery is subsequently moved to the patient’s left (Fig. 4). This procedure is ideally suited for patients with a non-intramural anomalous right coronary where the interarterial course typically runs somewhat above the base of the ‘V’ created by the two great arteries and thus creates space for the coronary by splaying the ‘V’ apart. The second pulmonary artery translocation operation is achieved by moving the pulmonary arteries anterior to the aorta (modified LeCompte manoeuvre). This procedure is our preference for treatment of a non-intramural anomalous left coronary, where the elongated LAD typically sits quite low and frequently has an intracanal course (Fig. 5). The anterior pulmonary artery translocation procedure effectively creates a 1 cm space at the base of ‘V’. The pulmonary artery translocation procedures, when applied appropriately, have proved to be efficacious in the treatment of patients with this specific form of AAOCA.

Numerous patients with AAOCA will also have abnormalities of the coronary arteries in a more distal location than the interarterial segment. Specifically, there have been 6 patients with a long, narrow intraconal left main coronary artery. Three patients had a myocardial bridge trapping the left anterior coronary artery that was identified preoperatively and confirmed intraoperatively. Both intraconal coronaries and myocardial bridges can be relieved by dividing the muscle fibres overlying the coronary artery. One additional patient had an occlusion of the proximal circumflex coronary artery. It was presumed that this was an acquired lesion, and was managed by placing a left internal mammary artery bypass graft to the distal circumflex in addition to unroofing the proximal coronary.

The indications for surgical repair of the patients in our study were quite diverse. Forty-one patients had preoperative symptoms of myocardial ischaemia, and 8 patients were asymptomatic but had an anomalous left coronary artery. These two indications are universally accepted indications for surgical repair [10]. There were 18 patients who had a variety of CHDs in association with AAOCA in whom the primary indication for surgery was the associated heart lesion. Finally, there was one set of siblings with AAOCA for whom the parents expressed their desire to proceed with surgery in the second child. This accounts for 68 of the 76 patients in this study. The 8 patients yet to be accounted for had an anomalous right coronary, did not have symptoms and did not have associated CHDs. We and others have suggested that patients with an intramural proximal coronary course should be added to the indication for AAOCA surgery, since these patients have a high incidence (85%) of developing symptoms of myocardial ischaemia [8, 9]. Therefore, there were a total of 4 patients in our current study who had an anomalous right coronary, did not have symptoms, did not have associated CHDs and did not have an intramural course. This subgroup, representing 5.3% of patients in our

Figure 5: (A) Illustration of the anterior PA translocation procedure. This procedure is ideally suited for an anomalous left coronary that originates from a single ostium, is not intramural and has an intraconal course. (B) The right PA is transected. The intracanal portion of the left main coronary is unroofed by dividing the overlying muscle fibres. (C) The right PA is moved anterior to the aorta, and is reanastomosed to the main PA (modified LeCompte manoeuvre).
study, may or may not prove to be a lower risk cohort. It is evident that these relatively rare subgroups of AAOCA will require additional studies to delineate the risk/benefit analysis of surgical intervention.

Several recent studies in the radiological literature have focused on the ability to accurately discern whether an anomalous coronary does or does not have an intramural proximal course [20–22]. This interest was stimulated by the observation that an intramural course may be an important factor in predicting the natural history of this disease [8, 9]. The ability of CT angiography to identify proximal intramural course in these studies has been quite modest. Thus, the important distinction between intramural and non-intramural course in asymptomatic patients with an anomalous right coronary currently cannot be made with definitive accuracy. It is also worth noting that while the prevalence of ischaemic symptoms is lower in non-intramural coronaries, half (4 of 8) of non-intramural right coronaries in our study were symptomatic. Because of this dilemma, we have recommended that all patients with AAOCA over the age of 10 years undergo surgical repair.

Paradoxically, one can make a reasonably accurate prediction concerning the intramural course solely based on a knowledge of the coronary artery patterns associated with AAOCA. In our study, 52 of 59 (88%) patients with AAOCA and 2 coronary ostia had an intramural course. Conversely, only 3 of the 17 (18%) patients with AAOCA and a single coronary ostium had an intramural course. Thus, an educated guess correctly predicts the intramural characteristics in 52 plus 14 = 66 of 76 patients or 87% of the time. Future studies regarding the prognostic capability of non-invasive modalities should at a minimum be held to the standard of an educated guess.

A separate controversy exists with regard to the need for follow-up studies and the activity limits on patients after AAOCA surgery [10]. Brothers et al. [11] observed that one-third of their postoperative patients had residual ‘ischaemic’ abnormalities detected using a combination of exercise stress testing, stress echocardiography and myocardial perfusion studies. However, it is unclear from their reports whether these patients had sustained myocardial damage preoperatively. They have recommended that all patients undergo stress testing postoperatively and have continued limitation of activity when abnormalities are detected. In our current study, we had 20 patients (fully half of all symptomatic patients) who sustained myocardial damage prior to their diagnosis. These patients most assuredly carry that damage with them postoperatively. Encouragingly, we have not had any patients develop new myocardial ischaemic events post-repair including the redevelopment of symptoms and/or a sudden death event. All of our patients undergo follow-up studies including echocardiograms and EKGs, but we have not implemented a systematic approach to stress-testing. We believe that the majority of postoperative AAOCA patients may return to unrestricted physical activities based on the absence of new ischaemic events in our patient cohort. However, an important caveat exists for patients who sustained myocardial damage preoperatively, as these patients are followed much more closely and are advised not to compete in high-level sports activities.

Nguyen et al. [23] recently reported the sudden cardiac death of an athlete just 2 months after repair of an anomalous left coronary. This would appear to raise a word of caution about the safety of AAOCA patients returning to physical activities. There are several details regarding this case that are important to highlight, including the fact that the patient had experienced a sudden death event and had sustained a myocardial infarction that was documented by the leakage of cardiac enzymes. It is conceivable that patients who have experienced sudden death should have a different threshold of testing and activity limits compared with other AAOCA patients.

In summary, this manuscript summarizes our experience with 76 patients who underwent surgical repair of AAOCA. The data highlight the diversity of the anatomic diagnosis of AAOCA and the complex surgical algorithm that is mandated to address the variety of forms of this disease. Based on an extensive postoperative observational experience, it is our conclusion that the proper application of this surgical algorithm results in a significant improvement in the intervened history compared with the natural history of this entity.

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REFERENCES

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APPENDIX. CONFERENCE DISCUSSION

Dr J. Hoerer (Munich, Germany): I have two questions. I agree completely with you that all these patients should undergo surgery; however, there is controversy with the guidelines in patients with anomalous right coronary and a non-intramural segment. So what type of diagnostics do you suggest to rule out whether there is an intramural segment of this right coronary artery? And based on your data, can you predict the possibility in those patients to have an intramural or no intramural segment?

Dr Mainwaring: It’s a good question. It really was a few years ago that we began focusing on whether these are intramural or not. So once that becomes something important, then the radiologists begin focusing on that. We have a pretty good radiology group at Stanford and yet they really have not been able to make this differentiation yet, but they’re working on that.

Now, you can make an uneducated guess at whether a coronary is intramural or not, because 72% are intramural, just taking the whole group, so you’re right three-quarters of the time. But you can make an educated guess and be even more accurate. We noted that in the patients who have 2 coronaries, 52 of the 59 patients were intramural and, conversely, in the patients who had a single coronary ostium, only 3 of 17 were intramural. So if you guess that the 2-coronary systems are intramural and you guess that the single coronary ostium are not, you are right 87% of the time. So going forward, the noninvasive diagnostic studies should be held to that standard. Seventy per cent isn’t good enough because that’s just random.

Dr Hoerer: And the next question, your surgical approach is quite convincing and straightforward. However, in your paper you reported on a few patients in whom you combined pulmonary translocation and unroofing. Unroofing of the intramural segment may be not sufficient because of a very short intramural distance or it may not be possible due to the subcommissural course of this intramural segment. Maybe you can comment on the technical aspects of this anomaly.

Dr Mainwaring: There is some variability in the intramural segment. We would combine the procedures when there is a very short intramural segment, like 1 or 2 mm, and you’re not convinced that’s going to relieve the potential for inter-arterial compression; then we would combine that with a pulmonary artery translocation procedure.

In the patients who have a low intramural course, one option is to take down the commissure, and the other option, if the intramural segment is long enough, is to fenestrate on the other side. There were about five or six patients that we fenestrated as opposed to taking down the commissure.

Dr M. Hazekamp (Leiden, Netherlands): Your presentation is quite supportive of my belief as a surgeon that the anomalous right coronary artery should also be addressed surgically. Our experience is much smaller than yours: we have 24 patients, but five of them had a near-death event prior to surgery and they were all aberrant right coronary arteries from that sinus. Now, all of them had intramural course. And I think that is very much supporting the evidence, the things that you say, that we should not be as conservative as the guidelines tell us. Of course, you’re also aware of this paper from Columbia in New York where they more or less say that as surgeons we should not be that aggressive. But I think we really should be aggressive because there is at least a chance of it, it’s difficult to prove that statistically, but even in the right coronary that is anomalously coming from the aortic root, there is a high chance of big problems. And the other thing that I wanted to comment on, if you push your radiologists really hard, they actually can say whether it’s an intramural course or not. If the slices are very, very thin, even I as a surgeon can see it on the CT scan, and that’s leading to a better indication probably.

Dr Mainwaring: I agree with you. I think the radiologists will be able to figure this out and be able to tell us quite accurately. It’s just that they didn’t understand until recently that this was something important.

Dr K. Francois (Gent, Belgium): I just want to ask you to specify the slit ostium: is that your intramural group, the ones who have really a slit ostium and it’s a very short intramural course, have you classified them within the intramural group?

Dr Mainwaring: We would call it intramural if it’s narrow at any point.

Dr B. Kreitmann (Marseille, France): Can you comment on a specific point? What kind of follow-up are you suggesting for these patients? It’s quite difficult to say that we have good results in patients that were asymptomatic and with no proof of ischaemia before. So that’s an interesting point.

Dr Mainwaring: Well, we now have over 400 patient-years of follow-up. And these patients have not had symptoms and they’re not having sudden death, so we feel pretty good about what we’re doing. One of the controversies that’s not answered is what can these patients do after surgery? We believe that patients who did not have evidence of myocardial damage are cured. So that’s three-quarters of the patients. So we let them do whatever they want. However, with the patients that have sustained myocardial damage we are more circumspect. We watch the patients who had a myocardial infarction very closely. Like adult patients that had an infarct, you have to let them heal that and then prove that they don’t have an arrhythmic site.

Dr S. Cicek (Istanbul, Turkey): We have a paediatric cardiologist here, Dr Eero Jokinen from Finland. What is your perspective, for example, for an asymptomatic, no ischaemia kid regarding treatment or follow-up?

Dr E. Jokinen (Helsinki, Finland): I’m just wondering how you find asymptomatic patients and find that he/she has a coronary anomaly as we don’t have any systematic screening programme. But anyway I agree that all teenagers who are identified as having a coronary anomaly and who are active in sports should have the operation done.