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Original Article

Primary care management of patients with common arterial trunk and transposition of the great arteries*

Jeffrey R. Boris

Division of Cardiology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America

Abstract Primary care cardiology, also known as ambulatory or outpatient cardiology, for the longitudinal management of patients with common arterial trunk or with transposition of the great arteries is both poorly described and has limited evidence to justify its basis. This article discusses some of the various complications that these patients can develop, reviews the medical literature, and describes a framework for care of these complex patients from infancy to transition to adult congenital cardiac specialist care.

Keywords: Ambulatory cardiology; outpatient cardiology; truncus arteriosus; transposition of the great vessels

Primary care cardiac management, also known as ambulatory or outpatient care, of patients with common arterial trunk or with transposition of the great arteries, has limited guideposts and few data. However, this is not unlike the care of patients with the large majority of congenital cardiac defects. Therefore, this paper will attempt to coalesce what few guidelines exist with some of the limited evidence and common-sense aspects of outpatient management of these patients in order to simplify and to give a framework for a standardised method of longitudinal outpatient management.

To date, the most comprehensive set of guidelines created for outpatient management of patients with complex congenital cardiac defects was published in 2006 from the Children's Hospital of Philadelphia.[1] This was a consensus-based document only, and was not based on data, nor was it subsequently validated. Despite these shortcomings, this is probably a reasonable place to start with the concepts of longitudinal care.

Another starting point is the consideration of the various presentations of patients with these defects. Patients with common arterial trunk can present as unrepaired or after surgical intervention, even years after birth. However, patients with transposition of the great arteries typically do not present without having had an intervention, as the incidence of survival past 1 year of age is extremely low. Patients with transposition of the great arteries can present within the 1st month or 2 of life without intervention, but the vast majority of patients will have undergone an atrial switch, such as a Senning or Mustard procedure, will have had an arterial switch, or will have had a more complex rerouting of blood flow, such as in those patients who have undergone a Rastelli operation.

Finally, the considerations for guidelines should take into account the type and frequency of testing performed in addition to obtaining historical and physical examination features on a routine basis in these patients. These three seemingly disparate parameters will be melded into what will hopefully be a reasonable guide to caring for these patients.

Common arterial trunk

The principles of management of patients in the clinic setting still revolve around the typical acquisition of a history and physical examination followed by measured utilisation of the various tools available to the cardiologist. Salient features of an appropriate history in any patient with cardiac disease are based upon ascertainment of evidence of

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Correspondence to: Dr J. R. Boris, MD, Clinical Associate Professor of Paediatrics, Division of Cardiology, Children's Hospital of Philadelphia, 3401 Civic Center Boulevard, Philadelphia, Pennsylvania 19104, United States of America. Tel: +1 215 590 3189; Fax: +1 267 426 5324; E-mail: borisj@email.chop.edu

the ability of the heart to meet the metabolic needs of the body. The questions posed to the family member or to the patient should be appropriate for the age of the patient. Thus, in infants with common arterial trunk who have not undergone surgical intervention, determination of the ability of the infant to eat without dyspnoea or diaphoresis is most important, although asking about cyanosis and syncope should be included in the routine features of the history, as well. In those older patients who have not undergone repair, but are at risk of developing severe vascular occlusive disease of the pulmonary tree and early death, inquiry into any limitations to their exercise, as well as history of oral hygiene, haemoptysis, sexual activity and contraception, and evidence of failure of the right ventricle, is essential. Features of the examination to assess include vital signs, such as weight, height, rate of respirations and pulse, and arterial saturations. The precordium may be active and may even demonstrate a tap secondary to hypertrophy of the right ventricle. There also may be a thrill felt over the outflow tract of the right ventricle. Auscultation of the heart can elicit a single heart sound, a click with ejection, a systolic ejection murmur, a diastolic murmur, or continuous murmurs associated with collateral vessels, either in the precordium or in the back. Assessment of the liver and the spleen may demonstrate organomegaly if there is failure of the right heart. Pulses may also be dynamic if there is any significant regurgitation of the aortic valve.

With advances in ability both to perform surgery and to improve recovery subsequently, patients with common arterial trunk are more frequently undergoing complete surgical repair in the neonatal period. However, this may vary with the ability of the individual centre; these centres may require the infant to wait until they are large enough to accept placement of a pulmonary homograft, or until they fail anticongestive medication therapy owing to worsening symptoms of congestive heart failure.

Regardless of the duration of the pre-operative period, it is vital to assess for evidence of other systemic defects or syndromes. DiGeorge syndrome, including those patients with deletion of chromosome 22q11, is frequently seen in patients with common arterial trunk. Associated defects can include interruption of the aortic arch, cleft palate, abnormal calcium homoeostasis, and abnormalities of the immune system. The manifestations of this syndrome can sometimes be subtle, and thus testing with fluorescent in-situ hybridisation for the chromosomal microdeletion or some other testing is routinely indicated for these patients. The vertebral–imperforate anus–cardiac–tracheo–oesophageal fistula–renal–limb (VACTERL) association also can have common arterial trunk associated with it, and, as such, should be suspected if there are other manifestations noted.

Once these patients have undergone their surgical intervention, their repair can be thought of as similar to that of tetralogy of Fallot. Early follow-up can be done for routine post-operative follow-up issues, such as those surrounding care of the wound and control of pain. This is often done within the 1st week after discharge from the hospital, assuming that there have been no complications. More complex follow-up evaluation, including echocardiography and electrocardiography, can likely start as early as 2–3 months after surgical repair, as the vast majority of healing is completed within this time. As per the guidelines published by Wernovsky and colleagues,[1] follow-up is done annually after that, including electrocardiography and echocardiography. Electrocardiography would be utilised for surveillance for evidence of sinus node dysfunction, as well as right ventricular hypertrophy, right axis deviation, and right bundle branch block. The right bundle branch block may even widen in conjunction with worsening right ventricular dilation and failure, similar to that in tetralogy of Fallot. Echocardiography would assess the right-sided path of blood flow, including right ventricular size, pressure, and function, pulmonary conduit stenosis and regurgitation, and branch pulmonary arteries. It could also assess left-sided structures, including neoaortic size, as well as residual aortic arch obstruction. Residual defects, such as ventricular septal defects or other residual shunts, could also be evaluated. Screening for abnormalities of behaviour and academic achievement should be assessed by 4–6 years of age, although consideration should be given to earlier screening if there are associated developmental concerns, especially in those children with the DiGeorge syndrome. Exercise stress testing should occur between ages 10 and 12 years and again between ages 14 and 16 years. In addition, cardiac magnetic resonance imaging can be performed between ages 10 and 12, although this would be most important if there was significant regurgitation of the pulmonary homograft for the evaluation of right ventricular volume and regurgitation fraction of the pulmonary valve, or if the distal aspects of the branches of the pulmonary arteries need further evaluation. Performance of Holter monitor testing for evidence of occult arrhythmias and sinus node dysfunction should likely be performed at least once per decade (Table 1).

Over longer follow-up, complications for which this surveillance is necessary can include residual ventricular septal defects, stenosis, and/or the aforementioned insufficiency of the pulmonary homograft, and stenosis of the branches of the pulmonary arteries. Chronic regurgitation of the pulmonary valve

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Table 1. Summary of outpatient management of patients with common arterial trunk after surgical repair.

- Infancy
  Post-operative
- Annual evaluation
  Including electrocardiogram and echocardiogram
- Age 4–6 years
  Screening for behavioural and academic difficulties
- Age 10–12 years
  Exercise stress testing
  Holter monitor
  Cardiac magnetic resonance imaging (as indicated for significant pulmonary regurgitation, for evaluation of right ventricular volume, for regurgitation fraction of the pulmonary valve, or for assessment of the distal portions of the branches of the pulmonary arteries)
- Age 14–16 years
  Exercise stress testing
  Holter monitor
- Transfer to Adult Congenital Heart Disease specialist
  Electrocardiogram, echocardiogram, Holter monitor, summary of operative course and outpatient testing

Adapted from Wernovsky et al[1]

can lead to dilation of the right ventricle with diminished systolic and diastolic function, arrhythmias, and intolerance of exercise effort. Furthermore, there can be residual regurgitation of the neoaortic (truncal) valve with or without dilation of the root of the aortic valve. The homograft will eventually require replacement, as the initially placed conduit as an infant will likely be too small to maintain adequate flow for an adult-sized heart, and typically undergoes changes that lead to worsening function of the valve.

As previously described above, patients with common arterial trunk who have not undergone repair eventually do have surgery in infancy. However, there are those rare patients who have enough balanced circulation and protection of the pulmonary circulation, such as those with severe stenosis or atresia of the main or branch pulmonary arteries, or those with only collateral circulation to the pulmonary arteries, who can survive undetected into childhood and even adolescence without intervention. These patients are at risk for pulmonary vascular occlusive disease and are at risk for extremely shortened lives. These patients need to be treated as those with Eisenmenger complex, including ensuring adequate hydration, appropriate immunisation, plus the use of contraception, iron, folate, and vitamin B-12 supplementation, prophylaxis for endocarditis plus appropriate oral hygiene, compression stockings for right ventricular failure, and pulmonary vasodilators, as appropriate.

Exercise capabilities in these patients deserve special mention. The 36th Bethesda Conference of the American College of Cardiology[2] specifically discussed aspects of competitive exercise germane to this class of patients, although leisure activities and exercise were not specifically addressed. In patients who have not undergone repair for their arterial trunk defect, they would be considered cyanotic. As such, the guidelines recommend participation in no greater than low-intensity, or Class IA, activities, as long as the following criteria are met:

- their arterial saturations remain .80%,
- they do not have tachyarrhythmias with exercise that can impair their consciousness, and
- they have no evidence of either moderate or severe dysfunction of their ventricles.

In these patients, it would be expected that they would have decreased chronotropic response, low physical capacity, and arrhythmias. There are no specific recommendations for those patients with common arterial trunk who have undergone surgical repair. Thus, it would be reasonable to again consider their defect as similar to that of tetralogy of Fallot. In these repaired patients, the guidelines recommend participation in any competitive activities as long as the estimated pressures in the right atrium and ventricle are "normal or near-normal", there is, at most, only mild right ventricular enlargement, there are no residual shunts, and there are no arrhythmias on either testing with Holter or exercise stress. However, if there is marked regurgitation of the pulmonary valve, pressures in the right ventricle are at or above 50% of the systemic arterial pressure, or there are arrhythmias, competitive exercise should be restricted to Class IA.

Transposition of the great arteries

Unlike patients with common arterial trunk, it is exceedingly rare for patients to present to clinic outside of the neonatal period with unrepaired arterial transposition.[3] Of those who present as neonates, they are likely to demonstrate cyanosis or evidence of congestive heart failure, reverse differential cyanosis – if in the presence of coarctation of the aorta – a gallop rhythm, or a murmur of either pulmonary valve stenosis or a ventricular septal defect. Although the vast majority of patients without repair of their arterial transposition do not survive past the age of 1 year,[3] those who do are likely to have a large defect of either the atrial or the ventricular septum, the left ventricle is likely to be de-trained, and they would probably need to undergo an atrial switch procedure, such as a Senning or a Mustard. The history in any patient without repair of their arterial transposition would include that of feeding efficacy and evidence of cyanosis. The examination can

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demonstrate a tap due to the right ventricle, a single second heart sound that is displaced from the normal parasternal location, and hepatomegaly, in addition to the aforementioned findings above.

Owing to the fact that survival of those patients with transposition of the great arteries without any repair is so rare, the remainder of this section will concentrate on those patients who have undergone some type of surgical intervention. In the present age, the vast majority of patients undergo an atrial switch, such as a Mustard or Senning procedure, or an arterial switch, such as an arterial switch or Rastelli. Similar to any cardiac defect, the surveillance depends upon the long-term known or expected complications associated with these surgical interventions. Thus, patients with an atrial switch can develop progressive failure of their right ventricle maintained in the systemic circulation, worsening regurgitation of the tricuspid valve, dysfunction of the sinus node, ventricular or atrial arrhythmias, obstruction of the atrial baffles, obstruction of subpulmonary outflow, and pulmonary hypertension. Therefore, in addition to the typical post-operative assessment, the Philadelphia guidelines[1] recommend the following testing every 1–3 years: electrocardiography, echocardiography, Holter, magnetic resonance imaging, and exercise stress testing. In addition, as indicated, cardiac catheterisation, ventilation/perfusion scanning, stress echocardiography, and B-type natriuretic peptide screening may be helpful (Table 2). The electrocardiogram and Holter monitoring would be expected to demonstrate the sinus node dysfunction and arrhythmias, with right axis deviation and right ventricular hypertrophy additionally shown on electrocardiogram. Echocardiography can be used to demonstrate flow across the atrial baffles, ventricular size, function, and estimated pressures, regurgitation of the atrioventricular valves, residual shunts, and assessment of the outflow tracts. Cardiac magnetic resonance imaging can demonstrate the baffle anatomy and flow more clearly if echocardiography is not able to elucidate this. Exercise findings noted in these patients often include decreased chronotropic response, decreased physical capacity, ST segment changes in the leads over the right precordium, arrhythmias, and desaturation.[4] Stress echocardiography and stress magnetic resonance imaging are investigational at this point, and have not been validated in this class of patients.[5] In these patients, the 36th Bethesda Conference recommends competitive exercise participation in low dynamic and low or moderate static sports (Classes IA and IIA) as long as there is no or only mild chamber enlargement on chest radiograph, echocardiogram, or magnetic resonance imaging, there are no atrial flutter, supraventricular tachycardia, or exercise-associated arrhythmias, there is no syncope or other cardiac symptoms, and there are normal results from exercise testing, including duration, workload, heart rate, electrocardiogram, and blood pressure response.[2] Some of these patients with impending failure of the right ventricle are considered for atrial switch takedown, with retraining of the left ventricle by placement of a pulmonary artery band followed by band removal and arterial switch.[6] This series of procedures has been recommended to be completed before mid-adolescence, as their outcomes are worse when they are older than age 12 years, meaning that patients who are to be considered for this pathway need to have this planned far in advance of adolescence.

Table 2. Summary of outpatient management of patients with transposition of the great arteries after atrial switch.

- Infancy
  - Post-operative
- Every 1–3 years
  - Electrocardiogram
  - Echocardiogram
  - Holter monitor
  - Cardiac magnetic resonance imaging
  - Exercise stress testing
- Age 4–6 years
  - Screening for behavioural and academic difficulties
- As indicated
  - Cardiac catheterisation
  - Ventilation/perfusion scan
  - Stress echocardiography
  - B-type natriuretic peptide
- Transfer to Adult Congenital Heart Disease specialist
  - Electrocardiogram, echocardiogram, Holter monitor, summary of operative course and outpatient testing

Adapted from Wernovsky et al[1]

In contrast, patients with arterial transposition who have undergone arterial switch are often felt to be more normal in both the short and long term in the physiologic and anatomic sense. Certainly, comparison of their historical features is similar after completion of their surgical procedures, including assessment of feeding and activity. However, their examination is definitely different as compared with those patients who have undergone either a Senning or a Mustard procedure. They can show systolic ejection murmurs, typically because of stenosis of the outflow of the right ventricle or of the Rastelli conduit. They can also demonstrate diastolic murmurs of regurgitation of either the pulmonary or the aortic valves. Their expected long-term complications include stenosis of the supravalvar region of the pulmonary artery, typically associated with the LeCompte manoeuvre or with narrowing along the suture line, or of the neoaorta.

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They can also have stenosis of the pulmonary arterial branches. The neoaorta can dilate, and its valve can become regurgitant. Similar to most patients who have had surgery for cardiac disease, the potential to develop dysfunction of the sinus node and arrhythmias exists and can worsen with time. They can demonstrate defects of coronary artery perfusion, which may be asymptomatic but can also demonstrate abnormal coronary flow reserve. Hypertrophy of bronchial collaterals is rarely seen, as is pulmonary arterial hypertension. In those patients who have had a Rastelli procedure, stenosis and/or insufficiency of the pulmonary conduit is a risk, as is subaortic obstruction of the baffle. Thus, surveillance evaluation is indicated on an annual or biennial basis, to include electrocardiography and echocardiography.[1] Similar to many other defects, academic and behavioural screening should be performed by 4–6 years of age. Exercise testing should be performed between ages 10 and 12 years and again between ages 14 and 16 years, with the latter test including myocardial perfusion at rest and with peak exercise. Magnetic resonance imaging of the heart can also be performed between 14 and 16 years if echocardiography is inadequate or if more information is required to assess the right ventricle. Finally, Holter monitoring probably should be performed at least once per decade. For those patients who are symptomatic or who may be considering highly competitive sports, consideration of cardiac catheterisation with angiography may be entertained (Table 3).

Table 3. Summary of outpatient management of patients with transposition of the great arteries after arterial switch.

- Infancy
  - Post-operative
- Every 1–2 years
  - Electrocardiogram
  - Echocardiogram
- Age 4–6 years
  - Screening for behavioural and academic difficulties
- Age 10–12 years
  - Exercise stress testing
  - Holter monitor
- Age 14–16 years
  - Exercise stress testing, including myocardial perfusion at rest and with peak exercise
  - Holter monitor
  - Consider cardiac magnetic resonance imaging if echocardiography is inadequate or more information is required to assess the right ventricle
- As indicated
  - Cardiac catheterisation, if symptomatic or if considering highly competitive sports
- Transfer to Adult Congenital Heart Disease specialist
  - Electrocardiogram, echocardiogram, Holter monitor, summary of operative course and outpatient testing

Adapted from Wernovsky et al[1]

Electrocardiography in these patients is often normal, although evidence of sinus node dysfunction can be uncovered. Echocardiography can be used to assess the outflow tracts for stenosis and valvular regurgitation; it can also demonstrate ventricular dysfunction and residual shunts. Holter monitoring can further delineate dysfunction of the sinus node, as well as arrhythmias of the atria or ventricles. They can also uncover an increased intrinsic heart rate, as these patients have been demonstrated to have increased sensitivity to the chronotropic effects of norepinephrine.[7] Interestingly, exercise stress testing has uncovered rhythm strips that suggest ischaemia, although the clinical correlation of this has not been defined.[4] Of note, myocardial perfusion testing has been variable in these patients. Coronary perfusion defects that are asymptomatic have been noted using sestamibi,[8,9] positron emission tomography,[10,11] and echocardiography utilising a dobutamine stress protocol.[12] However, completely normal studies have also been demonstrated with positron emission tomography in the absence of known ischaemia,[13] Doppler guidewire testing,[14] and magnetic resonance imaging using an adenosine stress protocol.[15] In all, much more study of the significance of coronary perfusion issues in those patients who have undergone arterial switch requires much more longitudinal investigation to be able to understand its significance.

For patients who have undergone arterial switch, participation in competitive exercise activities is completely unlimited as long as the function of their ventricles is normal, exercise testing is normal, and there are no significant arrhythmias.[2] Their competitive participation is limited to low dynamic and low or moderate static activities if they have more than mild haemodynamic abnormalities or ventricular dysfunction but as long as their exercise testing remains normal. It should be understood, however, that the Bethesda conference specifically addressed participation in competitive athletics. Thus, in distinction to competitive athletic recommendations, which are potentially restrictive, recreational athletics is increasingly being demonstrated to be helpful in the vast majority of patients with cardiac defects, in that it creates myocardial reserve, reduces the risk of obesity, assists in well-being and a feeling of normal from a psychological perspective, and improves mental performance in academic domains. Overall, patients with either common arterial trunk or arterial transposition should not be restricted from enjoyable participation in non-organised sports activities. However, they probably should not perform significant isometric activity if they have moderate or severe insufficiency of

the aortic valve or increasing dilation of the aortic root. In addition, they should likely not have exercise-induced ectopy. Consideration of the function of the systemic ventricle, especially if it is a right ventricle, may need to be factored into the degree of participation. Nevertheless, even patients with decreased ventricular function may be able to benefit from rehabilitation-oriented activities.

As has been previously mentioned, the degree of evidence-based data to guide the timing of follow-up and the utilisation of testing in that follow-up is limited and consensus based. Recently, the Children's Hospital of Boston has created a series of Standardized Clinical Assessment and Management Plans (SCAMPs), including one for the assessment of the outpatient management of patients who have undergone arterial switch with a history of arterial transposition or double-outlet right ventricle.[16] Their protocol includes outpatient visits at 1–3 weeks after surgical repair, which includes a chest radiograph, followed by visits at 6–12 weeks after repair, 6 months after repair, and 1 year after repair, the last of which includes a sedated echocardiogram. Subsequent visits occur biennially through age 18 years with an echocardiogram occurring each time. A lipid profile, which is a novel addition, is also performed at ages 6, 12, and 18 years. A stress echocardiogram or a radioisotope perfusion scan is performed at ages 12 and 18 years, as well. At this point, their specific Standardized Clinical Assessment and Management Plan protocol is in the phase of assessment, as they aggregate and assess their data. They expect that utilisation of resources will be higher in this earlier phase, but they expect a decrease in the utilisation of visits and echocardiograms pending further data analysis. Of note, Holter monitoring is not mentioned in their protocol, and thus outside of stress testing electrophysiology status is not considered here.

Prevention of endocarditis, which still remains a relevant concern despite the improvements in surgical outcomes, is widely known to have reduced need for antibiotic prophylaxis.[17] Thus, patients with common arterial trunk or arterial transposition with uncomplicated surgical repairs now no longer require specific antibiotic prophylaxis unless they are within the first 6 months after their surgical intervention, there are residual shunts near prosthetic material, there is prosthetic material that is known to have not endothelialised, or there is a history of prior infective endocarditis. Certainly, any unrepaired patient requires antibiotic prophylaxis due to persistent cyanosis. All patients should be encouraged to maintain good oral hygiene, to include routine dental evaluation.

As the population of these patients continues to age, the concepts of transition and transfer become more important. The ability to perform these two functions more completely and correctly has been better delineated with time.[18] Transition, as with any patient with complex congenital cardiac defect, needs to start when the patient is able to, at minimum, understand the fact that the child has a heart that is different. It may be helpful to use a question such as, "How much do you know about what is different about your heart?" Patients with either common arterial trunk or transposition of the great arteries have complex defects that require them to be followed up eventually by a specialist in adult congenital heart disease. At the time of transfer, the assurance that all summaries of operative and outpatient courses plus the latest electrocardiogram and echocardiogram are available for hand-off is one of the most important tasks that the paediatric cardiologist can accomplish.[1]

Similar to other defects, such as in hypoplastic left heart syndrome, specific data to guide ambulatory care beyond consensus statements are lacking.[19] However, the advent of the electronic medical record, as well as further efforts to collaborate and standardise data collection, continues to advance the ability to capture thse data. This will lead to standardised methods of routine assessment of these patients not dissimilar to a pre-flight checklist made by an airline pilot, with the electronic record providing the checklist. As humans, we cannot remember everything necessary, nor can we know at any one time all of the evidence that guides us towards best practices in the care of our patients. Guidelines such as these act as benchmarks and platforms to then launch us towards more appropriate and data-driven care of these complex children as they stretch towards adulthood. It is hoped that the contents of this article will be supplanted by more specific information in the future so as to understand further the array of complications seen in these patients and to better plan for their surveillance and management.

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