Cardiology in the Young (2014), **24**, 1057–1063
doi:10.1017/S1047951114001905

Original Article

Primary cardiovascular care for patients with valvar cardiac disease*

Jeffrey R. Boris

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

Abstract Outpatient management of patients with structurally or functionally abnormal cardiac valves combines an ability to record an appropriate history, a directed physical examination, a knowledge of both the interpretation as well as the context of any associated testing, and an understanding of the care guidelines in the medical literature. This article attempts to integrate these various features to guide the clinician towards more appropriate and timely management of patients with cardiac valve disease.

Keywords: Valvular insufficiency; stenosis; regurgitation; aortic root dilation

Received: 17 February 2014; Accepted: 17 February 2014

As has been mentioned elsewhere,[1,2] outpatient management of a large majority of children with cardiac defects is based on opinion and experience and very little data. There is not much difference in the care of those patients whose defects include abnormalities of their valves. Although the care of patients with this wide variety of anomalies has been addressed in occasional consensus documents, much of the research is derived from a combination of adult studies as well as observation. To be sure, the recommendations from the consensus documents as well as other papers have ventured further than those of the management of many other types of cardiac defects. Much remains to be determined, however, in the cementing of the aetiologies; the management strategies, both medical and surgical; and the long-term surveillance of this array of abnormalities which are diverse in severity, location, anatomy, and pathophysiology.

In starting with an approach towards primary care management, the patient must first have an assessment that includes the history and physical examination. As with a typical cardiac history, evaluation of the patient's response to increased cardiac activity is important. This includes dyspnoea, diaphoresis, cyanosis, syncope, chest pain, palpitations, and dizziness with exercise. As there is an association with certain types of valve diseases with specific genetic syndromes, assessment of this aspect of the patient's history is important as well. Often, clues to a patient's disease and prognosis can be augmented by questions regarding the family history. These include evidence of left-sided heart disease and connective tissue diseases, such as Marfan or Ehlers–Danlos syndromes, or a history of aortic dissection. Moreover, if there is a history of left-sided heart disease, ensuring that first-degree relatives are also evaluated is as important as the care of the patient.[3] The gestational history can further give aetiologic clues in certain valvular defects. For example, exposure to teratogens early in embryogenesis and cardiogenesis, such as lithium and benzodiazepines, has been associated with development of Ebstein's

*Presented at All Children's Hospital Johns Hopkins Medicine 14th International Symposium on Congenital Heart Disease, Saint Petersburg, Florida, 15–18 February 2014, Special Focus: Diseases of the Cardiac Valves from the Fetus to the Adult, Co-Sponsor: The American Association for Thoracic Surgery (AATS).

Correspondence to: Dr J. R. Boris, MD, Clinical Professor of Pediatrics, Children's Hospital of Philadelphia, 3401 Civic Center Blvd, Division of Cardiology Philadelphia, PA 19104, United States of America. Tel: +215-590-3180; Fax: +267-426-5324; E-mail: borisj@email.chop.edu

anomaly of the tricuspid valve. Recent data suggest that exposure to a variety of environmental organic compounds and metals, including benzene, butadiene, carbon disulfide, chloroform, ethylene oxide, hexachlorobenzene, tetrachloroethane, methanol, sulphur dioxide, toluene, lead, mercury, and cadmium, can lead to conotruncal malformations.[4] There had been prior research to indicate that embryonic exposure to selective serotonin reuptake inhibitor medications was also associated with valvar disease; however, subsequent data refuted this and demonstrated that these medications were safe to use from a foetal cardiac standpoint.

The physical examination still remains a very important part of the evaluation of the patient, as its correlation with non-invasive imaging gives the context in which the testing must be conducted. Inspection and palpation, though infrequently used, can still demonstrate evidence of ventricular volume loading in the face of semilunar valve insufficiency. A right parasternal lift, or heave, can indicate enlargement of the right ventricle, whereas a right parasternal tap suggests right ventricular hypertrophy. Determination of the placement of the point of maximal impulse, also known as the apical impulse, can give clues to left ventricular enlargement if it is leftwardly and/or downwardly displaced. However, auscultation is the most helpful aspect of the cardiac examination. The first heart sound, or S1, is lower pitched in tone, meaning that it is better heard with the bell of the stethoscope. It is best heard at the apex and lower border of the sternum. It can be decreased in amplitude in the regurgitation of the mitral valve or of the aortic valve. Conversely, it can have increased amplitude in mitral stenosis. The second heart sound, or S2, is higher pitched and best heard at the left middle border of the sternum, also known as Erb's point. It is better heard with the diaphragm of the stethoscope. Mildly fixed splitting of S2, while classically noted with atrial septal defects and right bundle branch block, can also be observed with mild stenosis of the pulmonary valve wide but mobile splitting of S2 is also seen in patients with mild stenosis of the pulmonary valve, plus in those with severe insufficiency of the mitral valve. Finally, although typically noted in patients with pulmonary hypertension, a single S2 is also encountered in patients with transposition of the great arteries as well as tetralogy of Fallot. The fourth heart sound, or S4, combines with S1 and S2 to make up one of the gallop rhythms. It is typically pathologic, although it can be seen in highly trained athletes and elderly people. It can often be found in patients with ventricular volume overload or dysfunction. However, for the purposes of this discussion, it can also be seen in patients with Ebstein's anomaly of the tricuspid valve, tricuspid atresia, aortic valve stenosis with severe left ventricular disease, and pulmonary valve stenosis with severe right ventricular disease. Clicks, which are snappy, high-pitched sounds typically because of turbulent vibrations downstream of an abnormal valve, are also usually pathologic. They routinely precede the murmurs of aortic and pulmonary valve stenosis, although they become less prominent as the stenosis worsens. They are also heard in patients with common arterial trunk, pulmonary valve atresia with ventricular septal defect, bicuspid aortic valve, prolapse of the mitral valve (as a mid-systolic click), and Ebstein's anomaly of the tricuspid valve (often occurring as multiple clicks). Of note, clicks do not occur in subvalvar or supravalvar stenotic lesions. Whoops, also known as honks, are loud, variable-intensity, and rather musical sounds. They accompany the murmur of regurgitation with prolapse of the mitral or tricuspid valve as well as the murmur of subaortic stenosis. They can evolve to become murmurs with time. Opening snaps, though not common in children, are routinely associated with mitral valve stenosis. A tumour plop, also rare in children, is a sound noted as a pedunculated tumour passes through an atrioventricular valve. Finally, murmurs, which are sounds associated with turbulence of the blood flow in the heart or great vessels, can occur in multiple circumstances involving the valves. They are seen in stenosis at the valvar, subvalvar, and supravalvar levels. Stenotic murmurs tend to be ejection in quality. They often get longer as the severity of the obstruction increases, although this can decrease as the ejection of the ventricle decreases with disease progression, such as with worsening stenosis of a semilunar valve. It can also be affiliated with an opening snap and diastolic rumble of atrioventricular valve stenosis. Regurgitant murmurs occur in mid to late systole, if associated with an atrioventricular valve, or in early diastole with a decrescendo quality, if noted with semilunar valve insufficiency.

Of course, evaluation for cardiac disease is not complete without an evaluation throughout the rest of the body for signs and symptoms of cardiac sufficiency or insufficiency as well as for other evidence of both the severity of the valvar lesions and of syndromes with cardiac as well as extra-cardiac manifestations. The latter includes evaluating and recognising specific facial features, skeletal findings, and other organ malformations seen in patients with various syndromes, chromosomal anomalies, and associations. An assessment of the lungs for tachypnoea, retractions, or congestion can suggest severe mitral valve regurgitation or stenosis. Finding abnormal situs of the liver can be associated with heterotaxy syndromes, which often include

atrioventricular septal defects and common atrioventricular valves. An enlarged liver can be seen in the face of severe tricuspid valve regurgitation, and a pulsatile liver is seen with either severe tricuspid valve regurgitation or elevated right ventricular pressure. Palpation of pulses can give plenty of important information; bounding pulses are seen in severe aortic valve insufficiency. Delayed and/or decreased femoral pulses are diagnostic of aortic coarctation, which is frequently seen in conjunction with bicuspid aortic valve. In addition, evaluation of jugular venous distension and pulsations can demonstrate evidence of tricuspid valve regurgitation or elevated right ventricular pressures. The extremities can also show signs of valvar disease, such as Quincke's pulses with aortic valve regurgitation, splinter haemorrhages in infectious endocarditis, and congenital abnormalities seen in various syndromes. Of course, no physical evaluation is complete without an evaluation of the vital signs, looking for tachycardia, tachypnoea, desaturation, a widened pulse pressure or a low blood pressure, and decreased weight, all of which can indicate various aspects of cardiac response to significant valvar disease.

Once valvar disease has been diagnosed or intervened upon with surgery or balloon valvuloplasty, routine surveillance in the clinic for evidence of progression of valvar disease or cardiac disease secondary to the valvar abnormality is paramount. Combining aspects of the history and physical examination with various tests help to determine the need for as well as the timing associated with intervention. Electrocardiography is commonly utilised as an adjunct screening test in the clinic, as it is relatively inexpensive, portable, and easy to perform. Such findings as the duration of the QRS interval as well as evidence for chamber enlargement can point towards secondary effects on the heart from valvular disease. Exercise stress testing can uncover further secondary effects on the heart, such as decreased exercise tolerance, ST and T-wave changes, and exercise-induced arrhythmias. The echocardiogram has become the test of choice, however, for paediatric cardiologists to get large amounts of data about the heart. It reveals both information about the diseased valve as well as the secondary effects on the heart, and does this as a non-invasive, mostly painless, and relatively portable test. The degree and progression of valve leak can be assessed through the peak and mean gradient across the valve, the width of the regurgitant jet, the regurgitant fraction, and evidence of flow reversal in either the aorta or the hepatic veins. Secondary effects on the heart can be determined through looking at the chamber size, such as end-diastolic and end-systolic dimensions of the ventricles, enlargement of the atria, or hypertrophy of the chambers, as well as by calculation of shortening fraction or ejection fraction plus measures of diastolic function. Finally, in neonates, revealing the presence or absence of a patent arterial duct can be helpful in supporting the systemic or pulmonary circulation, as needed. A technology that has progressed significantly over the last 15 years is cardiac magnetic resonance imaging. Although it is more expensive and more difficult to perform, the ability of magnetic resonance imaging to better evaluate the irregularly shaped right ventricle as well as to accurately gauge regurgitant fraction has improved the predictive ability of providers in the timing of intervention for valvular disease.

The next section of this paper will review specific valvular abnormalities and conditions, and discuss the indications for intervention. Much of the data presented are from the American College of Cardiology and American Heart Association Practice Guidelines for the Management of Patients with Valvular Heart Disease written in 2006.[5] Stenosis of the aortic valve is most frequently associated with a congenital defect in children, but can also be found owing to storage diseases and other metabolic disorders. It is one of the few defects in which good longitudinal data have been available for a long time, in the form of the Second Natural History Study.[6] In those patients with a peak-to-peak gradient determined by cardiac catheterisation to be >50 mmHg, morbidity was noted to be 1.2% per year, and sudden death was noted to be 0.3% per year. Therefore, intervention on the valve is felt to be appropriate with a peak gradient >64 mmHg or a mean gradient >40 mmHg. However, intervention can be considered with a peak gradient >50 mmHg or a mean gradient >30 mmHg in those patients who have symptoms, who participate in competitive athletics, who are pregnant, or who have electrocardiogram changes consistent with strain or ischaemia.

Aortic valve regurgitation can be due to a congenital anomaly of the aortic valve, or from an inflammatory cause, such as rheumatic fever, collagen-vascular disease, Kawasaki disease, or infectious endocarditis. Intervention is felt to be appropriate when the regurgitation is severe and chronic, and is associated with symptoms of syncope, angina, or dyspnoea on exertion. However, if the regurgitation is severe and chronic but the patient is asymptomatic, criteria of worsening function of the left ventricle, such as an ejection fraction <50% or left ventricular enlargement with a Z-score >+4, left-sided ST or T-wave changes on electrocardiogram, or concurrent aortic valve stenosis with a catheterisation gradient of >40 mmHg, intervention should be undertaken. Aortic valve prolapse is uncommon, and can occur in isolation or in

conjunction with a conoventricular/perimembranous ventricular septal defect. The Venturi effect created as the jet of blood crosses the ventricular septum from left to right can lead to chronic suction that deforms and eventually pulls down the right coronary cusp of the aortic valve. This deformation can be severe enough as to cause aortic valve insufficiency, but the indications for intervention should be no different than those listed above.

Bicuspid aortic valve is a common aetiology of aortic valve stenosis and regurgitation, as well as aortic root and ascending aortic dilation, in children and adolescents. It is considered to be the most common congenital cardiac defect, occurring in 0.5–2% of the general population. It can occur as an isolated finding, although it can also be seen in patients with coarctation of the aorta and in patients with Turner syndrome. As there are usually three leaflets in this semilunar valve, the bicuspid nature of the valve occurs when there is fusion of the raphe between two of the leaflets: this includes right–left fusion, right non-coronary cusp fusion, and left non-coronary cusp fusion. As more attention has been focused on this defect, increased data on the subtypes of the valve conformations have emerged. Right–left cusp fusion is the most common version of this valve, and is the type most frequently seen in patients with coarctation of the aorta. Right non-coronary cusp fusion, however, is typically associated with an increased incidence of aortic valve stenosis and insufficiency as well as aortic root dilation. Finally, left non-coronary cusp fusion is much less common than either of the other two conformations.[7,8]

Aortic root dilation has more recently been noted to be a complication of the bicuspid aortic valve as well as several other congenital defects. The greatest concern associated with aortic dilation is the ability for the aorta to dissect or to rupture. When seen in patients with common arterial trunk, tetralogy of Fallot, pulmonary atresia with ventricular septal defect, and transposition of the great arteries, it has felt to be a benign finding with little apparent risk of dissection, despite the high frequency of its appearance in these defects.[9] However, its association with bicuspid aortic valve, either in isolation or in patients with Turner syndrome, has definitely been seen lead to dissection and rupture, with sometimes fatal consequences. This dilation typically occurs at a younger age as compared with those patients with isolated aortic dilation without a bicuspid aortic valve, leading to the theory of the presence of an aortopathy in this entity. There has also been some recent thought that the type of cusp fusion in the bicuspid aortic valve can influence the type of dilation, either at the root or in the ascending aorta. This is only preliminary information, however, requiring further investigation. In patients with bicuspid aortic valve, intervention on the aortic root is indicated when the diameter of the aorta reaches >50 mm, or if the rate of growth of the diameter is >5 mm/year, or if the ratio of the cross-sectional area of the aorta to the height is >10 sq cm/m. If there is a need for concurrent replacement of the valve, intervention at a diameter of >45 mm or a cross-sectional area to height ratio of >8–9 sq cm/m is indicated.[5]

Aortic root dilation is also seen in patients with connective tissue disorders. These include Marfan syndrome, Ehlers–Danlos syndrome, familial thoracic aortic aneurysm and dissection, and Loeys–Dietz syndrome. Historically, Marfan syndrome has been the model for the management and intervention of aortic dilation, although it has become more widely recognised that these various disorders lead to different phenotypic outcomes in aortic dilation. Ehlers–Danlos syndrome has different subtypes, with the vascular, or formerly Type IV, variant associated with a much higher risk of aortic dilation and dissection. The classic and hypermobile subtypes reportedly have a 10% incidence of aortic dilation, although reports of dissection are quite rare in this subpopulation. Intervention for the aorta in Marfan, Ehlers–Danlos, and familial thoracic aortic aneurysm and dissection syndromes is felt to be indicated when the aortic diameter is >50 mm or at 45 mm when there is a family history of dissection at diameters of <50 mm. Loeys–Dietz syndrome has different indications for intervention, though; as it is associated with a much higher incidence of vascular dilation and rupture, the threshold diameter for surgical repair is 42 mm. In these patients, frequent surveillance with either magnetic resonance imaging or computed tomographic angiography of the vessels from the neck to pelvis is important,[10] although patients with Marfan syndrome should occasionally get more comprehensive imaging, as well as the entirety of their aortic arches can be progressively difficult to image as these patients age and grow.

Stenosis of the mitral valve can occur owing to a host of various congenital malformations, although it is more frequently noted after postoperative intervention or owing to the sequelae of acute rheumatic fever. When it occurs as a congenital defect, it can be isolated, or it can be associated with multiple left-sided lesions as part of Shone's complex, which classically includes a supravalvar stenosing mitral ring, a parachute mitral valve, subaortic stenosis, and coarctation of the aorta. This definition, however, has been broadened to include other types of left-sided obstructive lesions. The obstruction associated with mitral valve stenosis can cause left atrial enlargement and hypertension as well as pulmonary hypertension. Intervention is indicated when the mean estimated

Doppler gradient across the valve is >10 mmHg, regardless of the presence or absence of symptoms, or if the patient has the above gradient plus an estimated right ventricular pressure of >50 mmHg, as measured by tricuspid regurgitation jet, even if the patient is asymptomatic.[5]

Regurgitation of the mitral valve can also occur as a congenital finding, especially in association with an atrioventricular septal defect with common atrioventricular valve. Other aetiologies of mitral regurgitation include post-inflammatory causes, such as acute rheumatic fever, infective endocarditis, and other collagen-vascular disease, as well as storage diseases and connective tissue diseases, especially those associated with mitral valve prolapse. Of note, mitral valve prolapse can have atrial arrhythmias associated with it, which can cause patients to be symptomatic. Another potential aetiology of mitral valve regurgitation is systolic anterior motion of the anterior leaflet of the mitral valve. This is classically seen in patients with hypertrophic cardiomyopathy, but it can also be seen in patients after mitral valve repair, patients with hypertension, or in those undergoing stress testing using dobutamine as a chronotropic agent. The indications for intervention are severe regurgitation combined with the patient being symptomatic, or at the time of repair of the atrioventricular canal defect, or in an asymptomatic patient with severe mitral regurgitation plus an ejection fraction of <60%. Treatment of the underlying disorder is the therapy for systolic anterior motion of the anterior leaflet.[5]

Tricuspid valve regurgitation is most commonly associated with a congenital aetiology, especially with Ebstein's anomaly of the tricuspid valve. This can also have accompanying Wolff–Parkinson–White syndrome, acquired complete heart block, and pulmonary atresia, depending on the severity of the valvar leak. Other congenital aetiologies of tricuspid valve regurgitation include dysplasia of the valve, a cleft, a double-orifice valve, or in association with the common atrioventricular valve in atrioventricular septal defect. Acquired tricuspid valve insufficiency can happen in the setting of a shunt from the left ventricle across a ventricular septal defect to the right atrium, as well as with infective endocarditis, trauma, tricuspid valve prolapse, or acute rheumatic fever. Repair of the valve is indicated when there is severe regurgitation plus decreased exercise tolerance, or worsening systemic desaturation, or concomitant valve surgery on the left side of the heart. Intervention can be considered when there is associated atrial fibrillation, a cardiothoracic ratio >65%, or a post-traumatic flail leaflet.

In comparison, stenosis of the tricuspid valve is rare. It can be a congenital finding, but can also be seen after acute rheumatic fever, infective endocarditis, a tumour, or after surgical repair of a common atrioventricular valve. The indications for intervention include the presence of symptoms, such as fatigue, dyspnoea, and oedema, or concomitant surgery for a valve on the left side of the heart.

Stenosis of the pulmonary valve was the first congenital defect to undergo cardiac catheterisation with balloon valvuloplasty. It is most commonly a congenital finding, occurring either in isolation or with tetralogy of Fallot. It can also happen as part of dysplastic pulmonary valve syndrome, which can be isolated or seen in patients with Noonan syndrome. From the Natural History Study, mild stenosis tends to remain stable, or can even spontaneously improve or resolve.[11] Balloon valvuloplasty in the cardiac catheterisation laboratory is the routine method of reducing the gradient in these valves. Intervention should be undertaken if the patient is symptomatic, with findings such as dyspnoea on exertion, angina, or syncope, plus has an estimated peak gradient of >30 mmHg or if they are asymptomatic but have a peak gradient of >40 mmHg. Patients with dysplastic pulmonary valve syndrome can respond to balloon valvuloplasty, although most tend to require surgical intervention. There has been literature for several decades that debate the merits of balloon valvuloplasty in patients with tetralogy of Fallot at some point before complete surgical repair to reduce the need for placement of a transannular patch in the outflow tract of the right ventricle at the time of repair.

Unlike the congenital nature of pulmonary valve stenosis, and not including trivial physiological leaks noted on echocardiography, pulmonary valve regurgitation is usually acquired. It is most frequently seen after some type of intervention, such as placement of a transannular patch in patients with tetralogy of Fallot, although it can also be seen in valve-sparing attempts with resection of muscle bundles. It also can be seen after cardiac catheterisation with balloon valvuloplasty. An uncommon congenital cause of pulmonary valve insufficiency occurs in the setting of absent pulmonary valve syndrome, which can be an isolated finding or affiliated with tetralogy of Fallot. Rarely, connective tissue disorders can lead to pulmonary valve leakage. Indications for intervention for pulmonary valve regurgitation have changed over the last several decades. Historically, patients with tetralogy of Fallot who underwent placement of a transannular patch were felt to not need a competent pulmonary valve, with relief of the stenosis being paramount. As time went on, though, the realisation that these patients were significantly demonstrating early morbidity and mortality led to a shift in the approach towards these patients. At this time, the

indications for the symptomatic patient include the presence of right ventricular enlargement, right ventricular dysfunction, with an ejection fraction of <40%, the presence of ventricular arrhythmias, or congestive heart failure. For asymptomatic patients with right ventricular enlargement, a right ventricular volume of >150 ml/sq m, a regurgitant fraction of >40%, decreasing exercise tolerance, moderate or severe tricuspid valve regurgitation, or a QRS complex duration on the electrocardiogram of either >180 ms, or an increase of at least 3.5 ms/year are all indications for pulmonary valve intervention. These guidelines are in flux as more data are gathered in this cohort of patients, though.

One management concept notably absent so far from this discussion has been the use of medications to delay or to prevent the need for intervention, or to improve patients' symptoms. In all, there are little paediatric data to be able to assess efficacy. The use of β-blockers, angiotensin-converting enzyme inhibitors, and angiotensin receptor blockers are likely not helpful in aortic valve regurgitation. In fact, in adults, these are reserved for patients with either concurrent hypertension or left ventricular dysfunction. One class of patients in which β-blockade may be helpful is those with Marfan syndrome. These medications have been used for decades with notably mixed results. The Pediatric Heart Network is presently conducting a clinical trial comparing the use of atenolol versus losartan in patients with Marfan syndrome and aortic root dilation; results are expected within the next 1–2 years.[12] Diuretics can be used acutely in patients with new onset mitral or aortic valve regurgitation as well as potentially more chronically in patients with tricuspid regurgitation, although this is of some debate. They may also help in mitral valve stenosis. Digoxin has not been found to be beneficial in patients with significant valvular regurgitation. There are no medications that have been found to be helpful in patients with pulmonary valve regurgitation. The same could be said for those patients with valvular stenosis, although there have been controversial adult data to suggest that statin medications may be able to decrease certain types of progressive aortic valve stenosis.

One special class of medications deserves mention, which is the use of antibiotics for the prevention of infective endocarditis. Although the vast majority of valve disease previously required antibiotic prophylaxis at the time of various types of invasive procedures, these recommendations changed and diminished drastically in 2007.[13] The revised approach is the use of antibiotic prophylaxis only at times of dental cleaning or significant procedures that can lead to bleeding. It is no longer indicated for native valves with stenosis or regurgitation. In those with valve disorders, however, it is indicated for patients with prosthetic valves or valve material, patients who have undergone cardiac transplantation and have residual valvular regurgitation owing to a structurally abnormal valve, patients with a history of prior endocarditis, and patients with a congenital heart defect who have undergone intervention and have a residual defect at the site of a prosthetic device or material.

Overall, the primary care cardiac management of these patients involves a combination of knowledge of the historical and physical examination findings plus appropriate imaging and other testing as well as education of the patient and family to create a consistent model of surveillance that will catch what has previously been referred to as the "golden moment", that time that the patient is asymptomatic but has enough disease progression that further delay may lead to permanent damage to the heart or other morbidity for the patient. Thus, knowledge of the accompanying indications for intervention for these valves allow for further planning of this timing. One of the more difficult aspects of surveillance is determination of the timing of follow-up in these patients. A reasonable rule of thumb is that patients who are in the stages of their life of more rapid growth, such as infancy and adolescence, should probably be seen more frequently, as changes are likely to occur to the heart as rapidly. Those patients with more moderate or severe disease should be seen more frequently. Moreover, those patients who have remained stable can likely have their surveillance times lengthened; one method of estimating the revised time for follow-up is to double the time since the last visit in which the patient was seen. For example, if it had been 1 year since the last evaluation and there had been no progression or significant changes, stretching out to 2 years could be considered, with the understanding that the patient could be brought back into clinic sooner in the event of the onset of new symptoms or other clinical changes. It is expected that as time progresses and further data are collected in the management of paediatric cardiac patients, there will be an ability to have more evidence-based and protocol-based care. For now, however, maintaining an appropriate evaluation schema in combination with flexibility and an appropriate but questioning adherence to guidelines should serve these patients well.

Acknowledgements

None.

Financial Support

This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest

None.

References

1. Boris JR. Primary care management of patients with common arterial trunk and transposition of the great arteries. Cardiol Young 2012; 22: 761–767.
2. Boris JR. Primary care management of patients with hypoplastic left heart syndrome. Cardiol Young 2011; 21: 53–58.
3. Cripe L, Andelfinger G, Martin LJ, et al. Bicuspid aortic valve is heritable. J Am Coll Cardiol 2004; 44: 138–143.
4. Ngweze DP, Hornberger LH, Saretsky B, et al. Congenital heart disease and the emission of industrial developmental toxicants in Alberta, Canada. Circulation 2013; 128: A15332.
5. Bonow RO, Carabellow BA, Chatterjee K, et al. ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice guidelines (Writing Committee to Develop Guidelines for the Management of Patients with Valvular Heart Disease). J Am Coll Cardiol 2006; 48: e1–e148.
6. Keane JF, Driscoll DJ, Gersony WM, et al. Second natural history study of congenital heart defects. Results of treatment of patients with aortic valvar stenosis. Circulation 1993; 87: I16–I27.
7. Fernandes SM, Sanders SP, Khairy P, et al. Morphology of bicuspid aortic valve in children and adolescents. J Am Coll Cardiol 2004; 44: 1648–1651.
8. Russo CF, Cannata A, Lanfranconi M, et al. Is aortic wall degeneration related to bicuspid aortic valve anatomy in patients with valvular disease? J Thorac Cardiovasc Surg 2008; 136: 937–942.
9. Stulak JM, Dearani JA, Burkhart HM, et al. Does the dilated ascending aorta in an adult with congenital heart disease require intervention? J Thorac Cardiovasc Surg 2010; 140: S52–S57.
10. Hiratzka LF, Bakris GL, Beckman JA, et al. ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease. A report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologist, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. J Am Coll Cardiol 2010; 55: e27–e129.
11. Haves CJ, Gersony WM, Driscoll DJ, et al. Second natural history study of congenital heart defects. Results of treatment of patients with pulmonary valvar stenosis. Circulation 1993; 87: I28–I37.
12. Pediatric Heart Network: Marfan Study Basics. Retrieved 23 September 2014 from http://www.pediatricheartnetwork.org/Studies/CurrentStudies/MarfanStudy.aspx
13. Wilson W, Taubert KA, Gewitz M, et al. Prevention of infective endocarditis: guidelines from the American Heart Associatin: a guidelines from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Qualityof Care and Outcomes Research Interdisciplinary Working Group. Circulation 2007; 116: 1736–1754.