CARDIAC ISSUES IN THE ADOLESCENT AGE GROUP

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Heart disease in the adolescent age group has a unique spectrum. The previously unrecognized congenital heart disease may manifest for the first time in the second decade. These are usually milder forms of the congenital disease. Apart from this, the operated patient may manifest with new complications. These may include serious problems requiring re-operation (e.g., conduit stenosis) or may require prolonged therapy (e.g., oral anti-arrhythmics) or an intra-cardiac device implantation (e.g., automatic intra-cardiac defibrillator, stent implantation in post-op TOF). In addition, unoperated patients surviving into the second decade may have Eisenmanger's syndrome from left to right shunts. From cyanotic heart diseases left unoperated they may have severe cyanosis resulting in a hyperviscosity.

Acquired heart disease in the second decade is related to infections i.e. rheumatic heart disease, Kawasaki disease. Or they may be related to trauma (commotio cordis), drug abuse (severe hypertension, myocardial infarction) or extreme athletic training (LV hypertrophy-athletes heart). Systemic diseases (malignancy treated with chemotherapy; muscle dystrophy) may manifest with cardiac problems, which may limit their ultimate growth potential.

Key words: Adolescent, Heart disease, Arrhythmia, Complications.

THE adolescent age group is unique in several ways due to ongoing mental and physical development taking place and the behavior of teenagers during these years. This affects the differential diagnosis and management of medical conditions in this age group in general. The spectrum and presentation of cardiac disease during these years is also unique in many ways. In this article we will be discussing the spectrum of cardiac disease in the second decade of life and the variety of presentations it may have and focus on those potentially at high risk of mortality. *Table* 1 summarizes the groups under which one can categorize the presentations of heart disease. Each of these would be discussed individually including their differential and approach.

TABLE 1. ***Presentations of Heart Disease in Adolescent Years***

1.	Congenital structural heart disease: first presentation in the adolescent years
2.	Known unoperated heart disease presenting with complication
3.	Operated heart disease presenting with complication
4.	Heart disease acquired in the second decade
5.	Systemic disease manifesting with cardiac symptoms

Congenital Heart Disease: First presentation in adolescent years

Congenital heart disease may not always manifest with symptoms in the first few years of life. Problems such as atrial septal defect, mild form of Ebstein's disease or certain other conditions like a previously missed coarctation may manifest this late (*Figs.* 1a & 1b). Other conditions may not have been missed earlier but may *de novo* present in the second decade of life e.g., mitral valve prolapse, hypertrophic cardiomyopathy, corrected transposition, primary pulmonary hypertension. The usual presenting symptom is shortness of breath (ASD, Ebstein's) or may be palpitation or chest pain (MVP) or syncope (primary pulmonary hypertension). The management of individual conditions is guided by the hemodynamics and the course of the particular diagnosis.

The diagnosis is particularly challenging because the murmurs become more faint in the adolescent years due to the increasing chest wall thickness. Therefore the reliability on other modalities including X-ray, ECG and Echocardiogram increases. Studies evaluating the role of clinical exam and combination of investigations in patients with suspected cardiac disease in this age group have found echocardiograms to be most cost-effective and specific to make a conclusive diagnosis [1]. Occasionally the diagnosis may only be confirmed after a transesophageal echocardiogram (TEE).

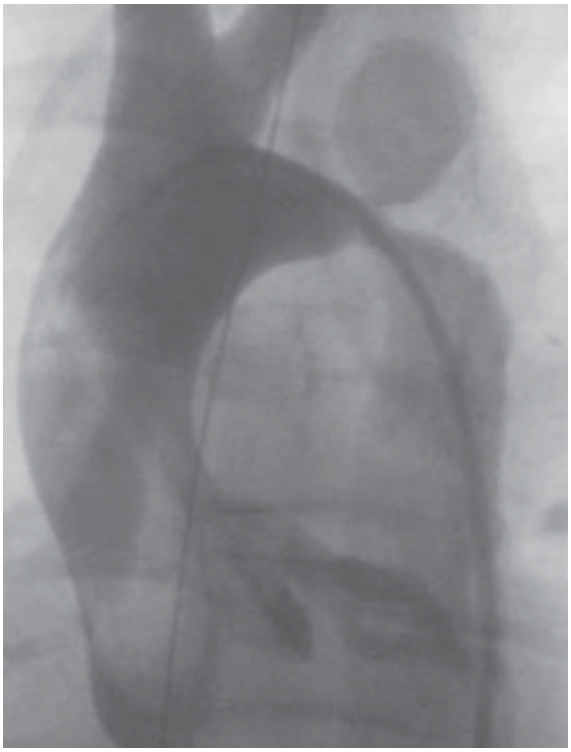

Fig. 1a. Panel on the left shows severe coarctation in an adolescent (14 yrs).

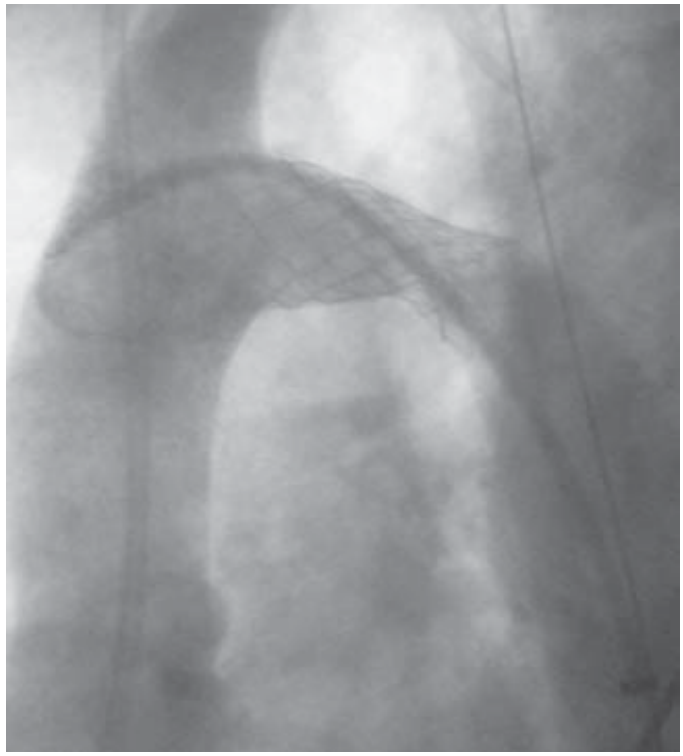

Fig. 1b. Panel on the right shows coarctation post-stenting.

It must be borne in mind that though any heart disease may present in the adolescent years, it would be unusual to see other than the ones mentioned above. The authors describe a 17-year-old male NYHA class I presenting with minimal symptoms was finally diagnosed to have Anomalous Left Coronary Artery from Pulmonary Artery (*Fig.* 2). The patient had normal cardiac function due to the collaterals.

Operated heart disease presenting with complication

These conditions would fall into certain categories and type of surgery undertaken in the first instance (*Table* 2). Infective endocarditis is one of the more common problems faced in an operated or previously known but unoperated congenital heart disease. In an operated patient the endocarditis maybe related to a residual lesion *e.g.* residual VSD or RVOT obstruction. For this reason the dental health and hygiene of these patients has to be habitually good.

Operated patient may manifest with arrhythmias [2]. The common problem includes a bradyarrhythmia related to previous surgery e.g., VSD closure, TOF repair or patients with complex heart disease including single ventricle. The bradycardia may be related to complete atrio-ventricular block or to junctional rhythm (*Table* 2). Tachyarrhythmias are more commonly related to atrial arrhythmia in the form of atrial flutter with or without high conduction rate to the ventricles: most often seen after Senning operation (Atrial Switch) or Fontan operation (palliation for single ventricle). One of the more common causes of ventricular tachycardia in a post-operative patient is the repaired Tetralogy of Fallot's with a ventriculotomy.

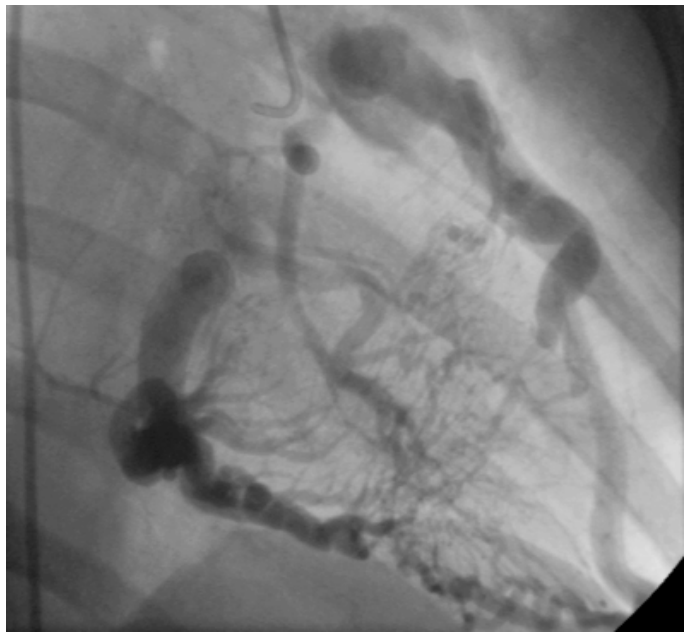

Fig. 2. A right coronary artery angiogram in a 20 yr old male with anomalous left coronary artery from the pulmonary artery. The left coronary artery is seen to be filling from the collaterals of the right coronary artery.

TABLE 2. ***Operated Heart Disease Presenting with Complications***

(i) Infective Endocarditis

(ii) Arrhythmia in a previously operated cardiac patient: Tachyarrhythmia
   1. Tetralogy of Fallot's
   2. Fontan operation
   3. Mustard, Senning, TAPVR

(iii) Arrhythmia in a previously operated cardiac patient: Bradyarrhythmia
   1. VSD closure
   2. Mustard, Senning, Fontan
   3. Tetralogy of Fallot's
   4. Complete Atrio-ventricular septal defect

(iv) Hypertension in previously repaired coarctation

(v) Myocardial infarction in a previous arterial switch patient

(vi) Pulmonary hypertension

(vii) Valve regurgitation
   1. Post-operative AV canal

(viii) Cardiomyopathy
   1. Post pump
   2. Secondary to residual lesion
      a. Pulmonary insufficiency in TOF
      b. AV valve regurgitation in AV canal
      c. Single ventricle palliation
         (i) Bidirectional Glenn shunt
         (ii) Fontan operation

Patients with a wide QRS (duration >0.18 ms) carry the highest risk of late ventricular arrhythmia. This ventricular arrhythmia is the cause of late mortality and sudden death in the postoperative TOF patients. The most common manifestation of an arrhythmia in a postoperative child is syncope followed by palpitation. Death may unfortunately be the first presentation of this type of ventricular arrhythmia. It is for this reason that the symptoms of syncope, pre-syncope and palpitation have to be investigated thoroughly and urgently including with an electrophysiology study. Usual treatment of this nature of ventricular tachycardia involves: (1) Correction of hemodynamically significant lesions *e.g.* pulmonary branch stenosis. (2) Medical management with anti-arrhythmics. (3) Medical devices like intracardiac defibrillators.

Unoperated congenital heart disease presenting with new complication in adolescent years

This would include a vast array of acyanotic and cyanotic heart diseases that have not been operated on either because they were thought to be insignificant or because the family decides against surgery (not an uncommon reason in the Indian medical practice).

Of the acyanotic heart diseases unoperated small VSD would be the most common CHD. These patients may present with a dilated left ventricle with mildly decreased LV function. The gradual volume overload of a small or a moderate VSD over the years may result in these findings. Other acyanotic heart diseases, which were left unoperated and present with fresh problems in this age group, include a large VSD, which has resulted in Eisenmanger's Syndrome. These patients at this stage may already be inoperable or may undergo evaluation for a high-risk surgical VSD closure.

The unoperated cyanotic CHD patients are typically TOF patients or are patients at the other end of spectrum of this disease i.e. Pulmonary Atresia with multiple aorto-pulmonary collaterals (MAPCA's). These groups of patients are presenting with complications of cyanosis, polycythemia and hyperviscosity syndrome. The complication of these may result in stroke, hyperuricemia and bleeding specifically from collaterals to the lungs (which may even be fatal) and require coiling (*Figs.* 3a & 3b). These patients may at this stage undergo an evaluation for operability. Other complications related to the hypercyanotic adolescents are mentioned in *Table* 3.

Acquired heart disease in the second decade of life (*Table* 4)

One of the more fulminant presentations of heart disease is

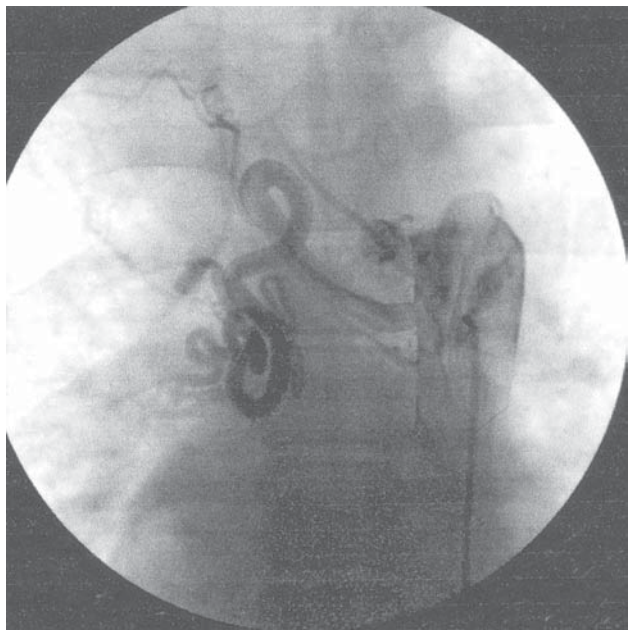

Fig.3a. Panel shows a large collateral in an unoperated cyanotic CHD. The collateral arises from the aorta and goes into the lung.

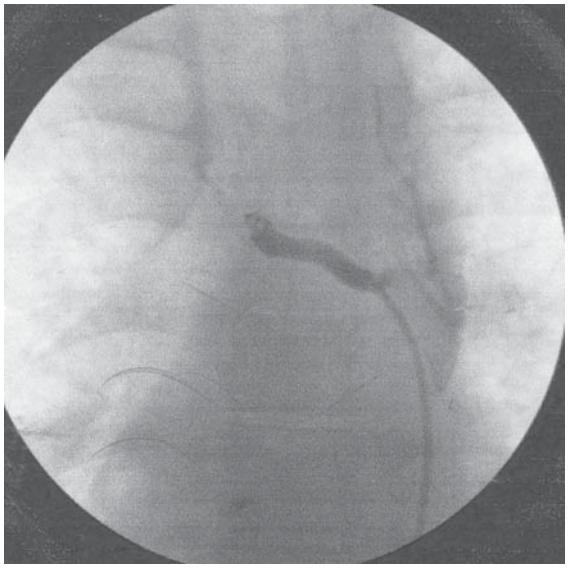

Fig. 3b. The panel shows the same post-coiling with no distal flow.

related to viral myocarditis. The patient may, within hours of being well, end up in the Intensive Care Unit and require inotropic support with high dose inotropes. These patients have a high mortality. On the other hand, myocarditis may also be present in a milder form, which may be difficult to recognize. This is the most common form where outcomes are better. The severity of illness is such that the recognition is usually easy with an X-ray showing cardiac enlargement or an echocardiogram showing decreased systolic cardiac function. The most specific test to diagnose this is Troponin I level which detects myocardial damage.

In addition to myocarditis, the patients in this age group may also have acquired heart disease in the form of Rheumatic fever or Kawasaki disease. The presentation of Rheumatic disease at this age is not uncommon though of Kawasaki disease, an already uncommon diagnosis would be relatively rare. The criteria for diagnosis would be similar to that at other ages and is currently beyond the scope of this article.

Acquired heart disease may also be related to abuse of drugs. The usual manifestations are related to acute intoxication of the drug and symptoms and signs vary to include hypotension (marijuana, narcotics) or hypertension (cocaine, phencyclidine, amphetamines), tachycardia (LSD), myocardial infarction (cocaine, amphetamines) or arrhythmia (amphetamines, volatile solvents, cocaine). In addition mainliners may have tricuspid valve endocarditis, which may have serious complications even requiring surgery.

"Athletes Heart" is a specific diagnosis where the left ventricular hypertrophy secondary to extreme exercise may result in difficulty in its differentiation from hypertrophic cardiomyopathy. The criteria to differentiate the two may require accurate measurement of the interventricular septal thickness, which should not exceed the measurement of 13 mm. If it does then the left ventricular mass index should be measured which should not exceed 134 g/m^2 in men or 110 g/m^2 in women. In addition, interventricular septum to free wall ratio of 1.3 or less would also confirm the hypertrophy to be physiological related to exercise. If these measurements are exceeded the patient would then be diagnosed to have true left ventricular hypertrophy. The increase in thickness of the wall may be concentric or eccentric. While anaerobic strength training produces concentric hypertrophy of the ventricular wall, endurance exercise on the other hand produces eccentric hypertrophy of the left ventricle [3].

TABLE 3. ***Complications of Unoperated Cyanotic Heart Disease in The Adolescent Years***

1. Hyperviscosity syndrome
2. Brain abscess
3. Stroke
4. Hyperuricemia
5. Hemoptysis
6. Severe hypoxia
7. Bleeding diathesis
8. Venous thrombosis
9. Failure to grow physically
10. Delayed puberty

TABLE 4. ***Acquired Heart Disease in Second Decade of Life***

1. Myocarditis
2. Drug abuse related heart disease
3. Athletes heart
4. Cardiac trauma
5. Kawasaki disease
6. Rheumatic heart disease
7. Takayasu's disease: Long segment coarctation; Branch Pulmonary Stenosis

Systemic diseases manifesting cardiac symptoms

Several systemic diseases including majority of the collagen vascular diseases may have cardiac manifestation. These include pericarditis or verucous endocarditis. The less common manifestations would include cardiac dysfunction secondary to myocarditis. Patients who have been treated for leukemia with Anthracyclines may manifest with cardiac

Key Messages

- Heart disease in Adolescent age group is varied and may be a manifestation of a previously undiagnosed congenital heart disease; a complication of a previously operated or unoperated heart disease.
- Previously operated heart disease patients may have varied complications including arrhythmia depending on the type of surgery. These need careful diagnosis and treatment as some of these maybe fatal.
- Specific problems may arise from vigorous athletic training resulting in severe LV hypertrophy. These need careful monitoring and evaluation since it potentially can result in sudden death.
- Other problems specific of this age include drug abuse related cardiac problems, trauma related cardiac injury, or cardiac manifestation of a systemic disease.

dysfunction, which may significantly alter the course and outcome of the disease inspite of being in remission. These patients would require routine cardiac follow up to detect the cardiac involvement early.

Similarly, patients with renal disease would have cardiac issue either in the form of hypertension or pericardial effusion.

Patients with neuromuscular disease including muscular dystrophy may manifest with cardiomyopathy in the second decade of life. The cardiomyopathy is usually the terminal event in these patients. Kearns-Sayre syndrome is a rare disease where, apart from neurological involvement, the cardiac conduction system is involved and results in a heart block.

The cardiac problems associated with thalassemia in the second decade of life are worth mentioning. Apart from myocardial iron deposition resulting in decreased myocardial function, the iron also gets deposited in the conduction system and results in ventricular arrhythmia.

Genetic syndromes like Marfan's and Ehler-Danlos may manifest with cardiac involvement, which may be serious and involve aortic dilatation, dissection or rupture requiring immediate surgery or resulting in death. The patients with Marfan's syndrome require regular assessment echo-cardiographically to assess aortic root dilatation is not progressive. Beta-blockers have been known to delay the progression of the aortic root dilatation and should be started in all Marfan's syndrome patients.

Conclusion

The presentation of cardiac disease in the adolescents is varied and differs in many ways from that of infants and children and even adults. They require a special perspective towards evaluation and management. Presentation of operated and unoperated congenital heart disease is different in adolescent age group. In addition, the complications of heart disease arising from previous surgery or the lack of it are unique too. Involvement of heart in systemic disease is significant and may even be the terminal event in these patients. Assessment of these patients by cardiologists at the right time is the key to appropriate evaluation and timely management.

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