Review Article

Manifestations of Pulmonary Disease in Adults with Congenital Heart Disease

Anup Kumar Singh[1], Nick Patel[2], Kevin Leung[3], Purvesh Patel[2] and Arunabh Talwar[2]

Internal Medicine, Unity Health System[1], Rochester, New York; and Division of Pulmonary, Critical Care and Sleep Medicine, North Shore-Long Island Jewish Health System[2], New Hyde Park, New York; and Department of Radiology, Nassau University Medical Center[3], East Meadow, New York, USA

ABSTRACT

Children with congenital heart disease (CHD) are more frequently living into adulthood as their survival has improved due to availability of better medical and surgical management in recent times. Management of adults with CHD is emerging as new challenge in the field of medical science. Adults surviving with CHD for longer duration have been observed to develop more complications as compared to children. It is important to recognise and treat these complications early to reduce the morbidity. Pulmonary diseases are the most common systemic complications associated with adults having CHD. These individuals are presenting to clinics or emergency for pulmonary complaints, hence, pulmonologist must be aware about the pulmonary manifestations of CHD and their management. **[Indian J Chest Dis Allied Sci 2013;55:85-95]**

Key words: Congenital heart disease; Adults; Pulmonary manifestations.

INTRODUCTION

Congenital heart disease (CHD) is one of the most common congenital defects and is estimated to occur in 6 to 8 per 1000 live births globally.[1-3] In India, the incidence of CHD has been observed to be 3.9/1000 live births and incidence of CHD is higher among pre-term as compared to full-term (22.69 *versus* 2.36 per 1000) live births.[4] Improvements in available diagnostic modalities, increased awareness among clinicians and better outcomes for corrective surgery have resulted in prolonged survival of children with CHD.[5,6] It has been estimated that more adults than children are living with CHD and this has been increasing at the rate of 5% per year.[5] In the United States, extrapolating this rate, it is estimated that there are more than one million adults with CHD.[5] The advancing age of this population, however, has been associated with the development of complications that clinicians were not seeing often. Proper identification and management of these complications would significantly impact the survival and quality of life of adults with CHD. By far, the majority of these are pulmonary complications and these significantly contribute to morbidity and mortality. This review provides an overview regarding the various manifestations of pulmonary disease in adults with CHD.

ADULT CONGENITAL HEART DISEASE

The prevalence of congenital heart disease (CHD) in adults has been observed to be 4.09 per 1000. The prevalence of adult CHD is more in females (4.55/1000) as compared to males (3.61/1000).[7] There is no gender difference in mortality in adults with CHD but the risk of cardiac outcome (pulmonary hypertension, infective endocarditis) varies with gender.[8] Atrial septal defect (ASD) with a prevalence of 0.88 per 1000 adults and ventricular septal defect (VSD) with a prevalence of 0.78 per 1000 adults are the most common non-severe forms of CHD in adults. Tetralogy of Fallot (TOF)/truncus arteriorus (conotruncal anomalies), artio-ventricular cushion defects, univentricular heart, transposition complex are categorised under severe CHD with an overall prevalence of 0.38 per 1000 adults. Conotruncal anomalies and endocardial cushion defects are the most common lesions among severe CHD.[7] Non-severe CHD are of importance for the pulmonologist as these patients have more chances of developing pulmonary complications due to prolonged survival.

Limited data are available regarding the prevalence of adult CHD from India. ASD, TOF and coarctation of aorta (CoA) account for 50% of the patients requiring surgical treatment in adulthood.[9] The number of adult patients with ASD requiring

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Correspondence and reprint requests: Dr Arunabh Talwar, North Shore-Long Island Jewish Health System, Division of Pulmonary, Critical Care and Sleep Medicine, 410 Lakeville Road, Suite 107, New Hyde Park, NY 11040, USA; Phone: (516) 465-5400; Fax: (516) 465-5454; E-mail: Arunabh@nshs.edu

surgery has decreased but there has been an increase in the number of patients with TOF, CoA and other complex lesions requiring surgery in recent years among Indian population.

MANIFESTATIONS OF PULMONARY DISEASE IN ADULT CONGENITAL HEART DISEASE

Pulmonary disorders and adult CHD are associated with each other as components of either CHD syndromes or connective tissue diseases (CTDs).[10] These may also manifest as complications of CHD itself or surgical procedures for CHD. Congenital heart disease syndromes, like Turner's syndrome, William's syndrome, lentigines, electrocardiogram conduction abnormalities, ocular hypertelorism, pulmonic stenosis, abnormal genitalia, retardation of growth, and sensorineural deafness (LEOPARD) syndrome, Down's syndrome, Noonan syndrome and Scimitar syndrome have pulmonary manifestations. Most common CTDs associated with both adult CHD and pulmonary diseases are Marfan's syndrome, Ehlers-Danlos syndrome (EDS), and osteogenesis imperfecta (OI). These pulmonary manifestations can be divided into different groups (Table 1).

Table 1. Manifestations of pulmonary diseases associated with adult congenital heart disease

Chest Wall Disorders
Pectus excavatum
Pectus carinatum
Kyphoscoliosis
Straight back syndrome
Shield chest
Parenchymal and Airway Disorders
Congenital lobar empysema
Pulmonary hypoplasia
Bronchiectasis
Pulmonary Vascular Disorders
Pulmonary arterial hypertension
Pulmonary thromboembolism
Pulmonary vascular anomalies
Pleural Disorders
Pneumothorax
Pleural effusion
Obesity Associated Pulmonary Manifestation
Pulmonary Function Limitations

Chest Wall Disorders

Chest wall disorders are observed in 1% of general population.[11] Congenital deformities of chest wall are present in 1 in 300 individuals.[12] Chest wall abnormalities are seen in 0.17%[13] individuals with adult CHD; most often as a part of a congenital syndrome or hereditary disorder. Anterior chest wall deformities (*pectus excavatum and pectus carinatum*) are predominant chest wall disorders.

Pectus Excavatum

Pectus excavatum is defined as a concave depression in the lower part of the anterior chest wall. The deformity is caused by the over-growth of ribs pushing the sternum into a posterior position.[14,15] It can be detected more easily during the growth phase in puberty.[12] *Pectus excavatum* is the most common chest wall deformity accounting for more than 85% of chest wall disorders and is present in 1 out of 400 children.[16] It is more common in males at a ratio of 5:1; 40% of individuals will have a family member with a similar deformity.[17]

Pectus excavatum has been associated with Marfan's syndrome, EDS and congenital cardiac anomalies like mitral valve prolapse (MVP)[7,8] and ASD. MVP is the most common CHD present in 16% cases with *pectus excvatum*, followed by Marfan's syndrome (3.1%) and EDS (2.1%).[18] *Pectus excavatum* is more severe in patients with Marfan's syndrome and should be ruled out in all patients with *pectus excavatum* and scoliosis. In patients with Marfan's syndrome, assessment with echocardiography is important to detect associated CHD, such as, aortic root dilatation, aortic valve incompetence and MVP.[19] Congenital heart lesions associated with EDS include ASD, VSD, TOF, bicuspid aortic valve and persistent atrioventricular canal.[20]

Patients with *pectus excavatum* complain of dyspnoea on exertion, chest pain, palpitations, frequent chest infections and often develop exercise-induced asthma.[21] Pulmonary function test (PFT) will reveal a mild restrictive pattern with total lung capacity (TLC) and forced vital capacity (FVC) in the low-to-normal range.[22] The degree of concavity is considered significant when the distance between the surface of the anterior wall of the thorax and the deepest part of the depression is greater than 3 centimeters on a lateral chest radiograph (Figure 1).[23] Thoracic computed tomography (CT) is useful for operative assessment. CT of the chest facilitates calculation of Haller's index which is computed by dividing the transverse diameter by the anterior-posterior diameter at the deepest level of depression. A Haller's index of greater than 3.25 is considered favourable for surgery.[24] However, exercise and physiotherapy are also acceptable treatments as many people lead normal lives without surgical interventions. Some studies[25,26] have reported a deterioration in pulmonary function following surgery, which suggests that improvements in dyspnoea, exercise tolerance and chest pain after surgical repair are more likely to be related to cosmetic changes.

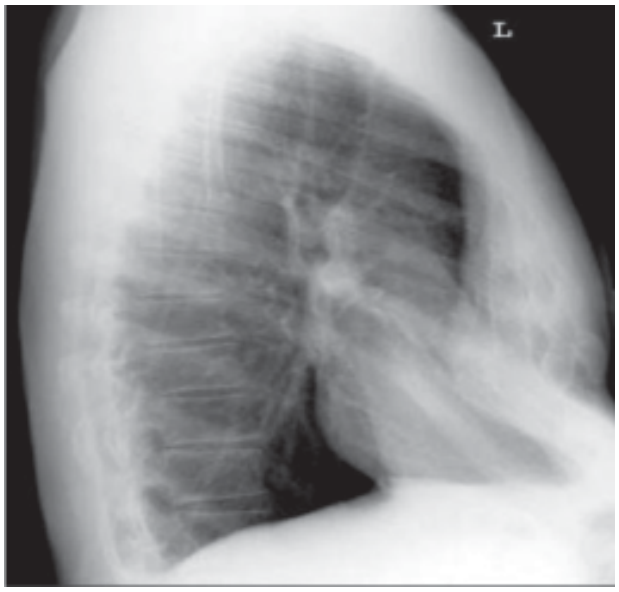

Figure 1. Chest radiograph (lateral view) showing depression of the lower end of sternum suggestive of *pectus excavatum*.

Pectus Carinatum

Pectus carinatum is characterised by protrusion abnormalities of the anterior chest wall (Figure 2). *Pectus carinatum* is caused by an abnormal growth of costal cartilage which pushes the sternum forward. The deformity is rarely noticed at birth and accentuated during the growth phase of adolescence reaching its peak in adult life. It is present in 1 out of 1000 adolescences and constitutes 5%-15% of chest wall disorder.[27,28] It is more common in males (male:female=4:1) and 26% of patients report a family history of the condition.[28,29]

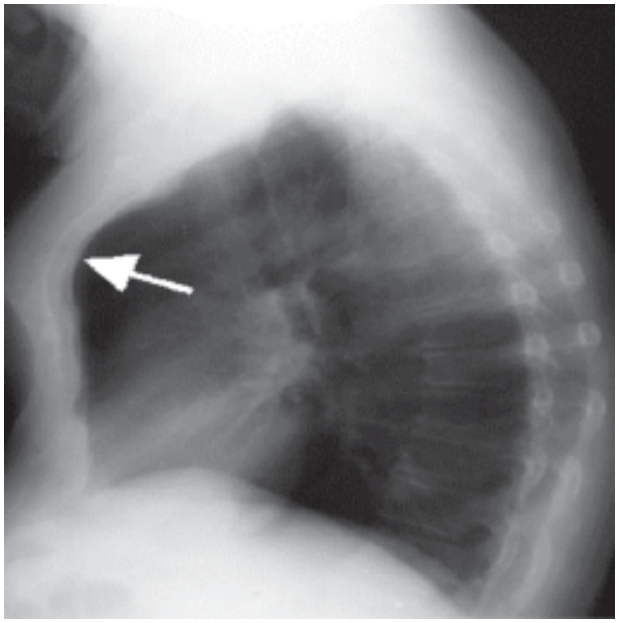

Figure 2. Chest radiograph (lateral view) showing protrusion deformity of sternum (arrow) suggestive of *pectus carinatum*.

VSD is the most common CHD associated with *pectus carinatum* followed by patent ductus arteriosus (PDA), ASD, TOF, MVP, mitral stenosis and CoA.[30,31] Marfan's syndrome and its associated congenital heart lesions (like, aortic root dilatation, aortic valve incompetence and MVP) are associated with 15% cases of *pectus carinatum*.[28] Patients report palpitations, dyspnoea, and wheezing that are accentuated during exercise. Conservative treatment like physical and orthopaedic therapy are not useful as most symptoms can be attributed to cosmetic defect of the chest wall. In the absence of accompanying diseases these symptoms should disappear after surgery.[26] Surgical treatment principally involves subperichondrial resection of the cartilages and sternal osteotomy.[32]

Kyphoscoliosis

Kyphoscoliosis is a vertebral column defect characterised by both lateral (scoliosis) and antero-posterior (kyphosis) curvature of the spine (Figure 3). Kyphoscoliosis can be congenital, idiopathic or secondary to vertebral destruction. Congenital and idiopathic kyphoscoliosis is due to defects in the development of vertebrae and hereditary in origin. It is associated with Marfan's syndrome (MVP, aortic dilatation, AV incompetence), adults with William's syndrome (supravalvular aortic stenosis), pulmonary stenosis (PS), mitral regurgitation (MR) and Friedriech's ataxia (hypertrophic cardiomyopathy).[33-35]

Severity is assessed by measuring the Cobb angle (the angle formed by the intersection of two lines, each of which is parallel to the top and bottom vertebrae of the scoliotic or kyphotic curves).[36] The deformity is considered severe with a very high risk of respiratory

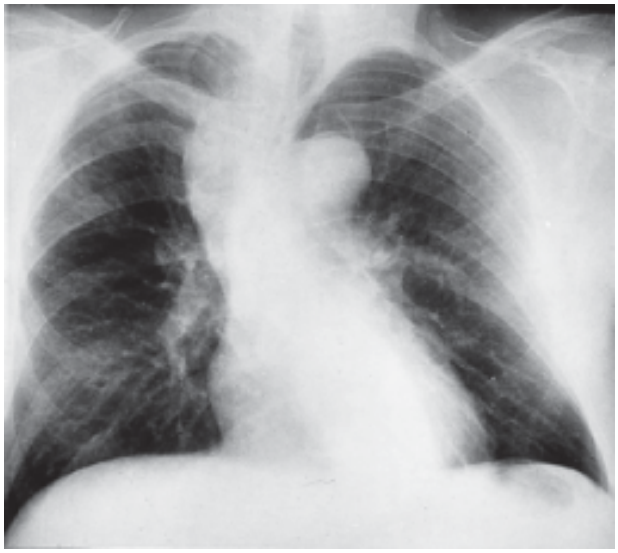

Figure 3. Chest radiograph (postero-anterior view) showing bulging of the right mediastinal outline caused by kyphoscoliosis. Intercostal space narrowing on the left as compared to the right side is also seen.

failure at Cobb angle greater than 100.[37] Alveolar hypoventilation, sleep disordered breathing, pulmonary hypertension and cor-pulmonale are common pulmonary complications in patients with kyphoscoliosis.[38] TLC, FVC and functional residual capacity (FRC) decrease with no change in residual volume (RV).[39] Physiotherapy and oxygen administration are useful supportive measures. Non-invasive positive pressure ventilation (NIPPV) is effective for chronic alveolar hypoventilation. Patients with kyphoscoliosis respond to NIPPV with improved gas exchange, quality of sleep, day-time function and reduction in hospitalisations.[40] Surgical procedures like spinal fusion or insertion of Harrington rods are important in patients with kyphoscoliosis secondary to neurological disorders to prevent progressive myelopathy.

Straight Back Syndrome

Straight back syndrome results due to congenital failure of development of the normal adult kyphotic curve. A patient is said to have straight back syndrome when the ratio of the transverse to antero-posterior diameter of greater than 3 and loss of normal dorsal kyphosis.[41] This syndrome is also known as "pseudo-heart disease syndrome", as reduced antero-posterior diameter compresses the heart and great vessels between the sternum and vertebrae. Hence, the heart appears enlarged on chest radiograph and auscultation may reveal a murmur in the absence of cardiac defects (Figure 4).[41] Lateral chest radiograph, echocardiography and cardiac catheterisation can help to rule out cardiac disorders. Although this entity has no association with CHD but it is worth mentioning as several of these patients can get be wrongly labelled as having CHD. It is also important to recognise this syndrome to avoid false diagnosis during employment and with insurance related issues.

Shield Chest

Shield chest is a broad shaped chest with widely shaped nipples and an increased angle between the manubrium and the body of sternum. Poor development of lymphatic channels during foetal life is responsible for shield chest.[42] This condition is common in Turner's syndrome (biscupid aortic valve, CoA)[42] and LEOPARD syndrome.[43] Echocardiography is technically difficult in these patients and should be done by the experts only. These patients do not have any functional impairment, and hence, shield chest is more of a cosmetic defect.

Parenchymal and Airway Disorders

Parenchymal and airway disorders may be congenital in origin or secondary to CHD. These can be further classified into different groups as described here.

Congenital Lobar Emphysema

Congenital lobar emphysema (CLE) involves hyperinflation of a lobe of the lung with compression of normal lung parenchyma and contralateral displacement of the mediastinum. It is a rare congenital malformation with a prevalence of 1:20000 to 1:30000.[44] Males are affected more than females, in a ratio of 3:1.[45] Aetiology of CLE is multifactorial. Obstruction to the developing airway (which may be intrinsic or extrinsic), leading to ball-valve mechanism and resultant air trapping and emphysema is considered to be the possible underlying mechanism. Left upper lobe is most commonly affected followed by the right upper and

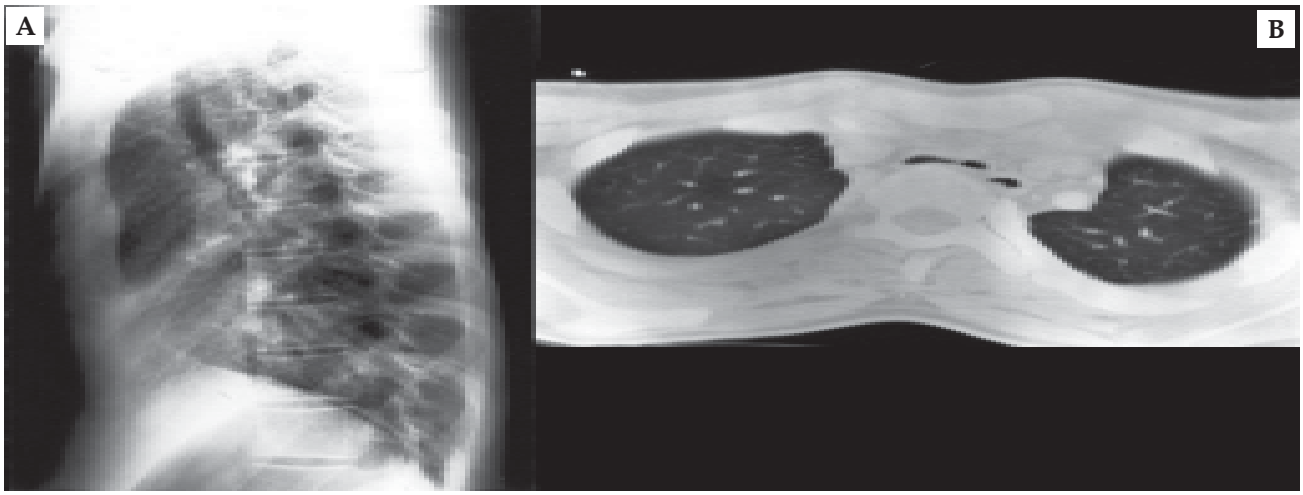

Figure 4. Chest radiograph (lateral view) (A) showing loss of thoracic kyphosis; CT chest (lung window; at the thoracic inlet) (B) of the same patient showing trachea is flattened to a slit between the manubrium sterni and the vertebral column. These findings are classically observed in "straight back syndrome".

middle lobes; but, any lobe may be affected.[46] Lobar emphysema in cases with CHD is caused by partial obstruction of bronchi by an enlarged pulmonary artery or left atrium.[47] Almost all cases of CLE are diagnosed before the age of six months, and hence, very few cases have been diagnosed directly in adulthood without any symptoms in childhood.[47] About 12%-14% of CLE cases are associated with CHD.[45] Among acyanotic CHD associated with CLE and VSD is most common (41.7%), followed by PDA (27.8%).[48] TOF with absence of pulmonary valve leaflets is the most common cyanotic CHD described in association with CLE.[48]

Individuals with CLE have frequent respiratory infections, dyspnoea, cough, wheeze and cyanosis. Chest radiograph and CT are diagnostic and show the hyperluccent affected lobe with herniation to the opposite side, shifting of the mediastinum to the opposite side and collapse of the remaining ipsilateral lung (Figure 5).[49] Lobectomy has been considered to be the treatment of choice in CLE. However, the occurrence of this condition in many asymptomatic individuals has prompted a shift from surgical to conservative management.[50]

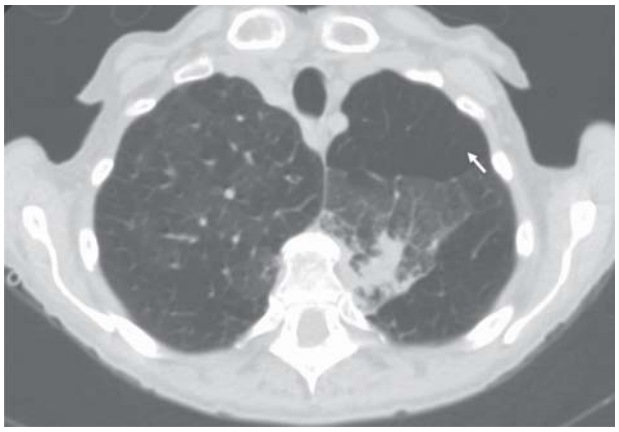

Figure 5. Computed tomography of chest (lung window) showing localised hyperlucency anteriorly (arrow) on the left side suggestive of congenital lobar emphysema.

Pulmonary Hypoplasia

Pulmonary hypoplasia is characterised by incomplete development of distal lung parenchyma. Pulmonary hypoplasia can be primary or secondary to congenital diaphragmatic hernia, oligohydroamnios and CHD. Primary pulmonary hypoplasia has mortality of around 71%-90%,[51] and hence, its presentation in adults is not very frequent. Secondary pulmonary hypoplasia is more common in adults as compared to primary pulmonary hypoplasia. Secondary pulmonary hypoplasia in patients with CHD develops due to reduced pulmonary blood flow as severe congenital pulmonic stenosis leads to pulmonic hypoplasia in adults.[52] Total or partial anomalous pulmonary venous connection (TAPVC) of the right lung to the inferior vena cava and right pulmonary artery hypoplasia in adult form of *Scimitar* syndrome are associated with pulmonary hypoplasia.[53]

Adults with pulmonary hypoplasia may be asymptomatic or present with frequent respiratory infections or pulmonary hypertension. Diagnosis of pulmonary hypoplasia is facilitated by imaging. Evaluation of the degree of hypoplasia requires lung scintigraphy and magnetic resonance imaging (MRI). Cardiac lesions are evaluated by echocardiography and Scimitar syndrome is diagnosed by pulmonary angiography. Early surgical repair of CHD leads to normal growth of the lungs, and hence, prevents the development of secondary pulmonary hypoplasia in adults.[54]

Bronchiectasis

Bronchiectasis is an abnormal permanent dilatation of the proximal airways (Figure 6). Bronchiectasis and CHD co-exist in individuals with primary ciliary dyskinesia (PCD). Bronchiectasis is present in all adults and 50% of children with PCD.[55] The prevalence of CHD with heterotaxy is 200-fold higher in patients with PCD than in the general population. Cardiac anomalies commonly present in individuals with PCD are inferior vena cava drainage via azygous, TGA, VSD, ASD and subpulmonic stenosis.[56] Bronchiectasis must be ruled out as a cause of frequent respiratory infection in adults with CHD. Bronchiectasis has also been associated with TAPVC drainage in few cases of Turner's syndrome.[57]

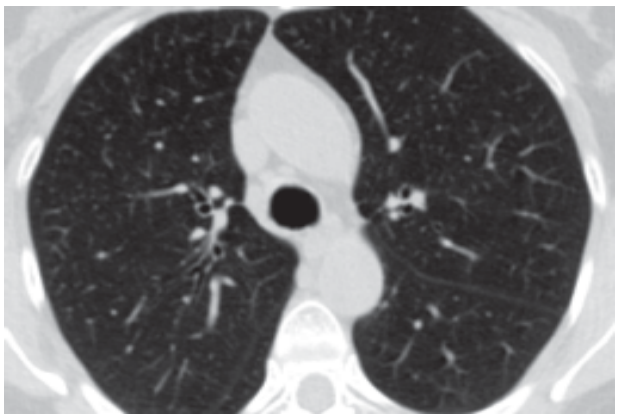

Figure 6. Computed tomography of chest (lung window) showing central bronchiectasis.

Pulmonary Vascular Disorders

Pulmonary hypertension is the final outcome of all pulmonary vascular disorders. Pulmonary hypertension may be a manifestation of complex pathophysiology of adult CHD or it may be caused by increased pressure in various congenital pulmonary vascular anomalies. Pulmonary vascular disease

secondary to CHD is a preventable illness, especially, if repair is offered to children under two years of age.[58] This can be further divided into different groups as described here.

Pulmonary Hypertension

Pulmonary hypertension is defined as a mean pulmonary artery pressure greater than 25mmHg at rest, pulmonary capillary wedge pressure greater than 15mmHg, and pulmonary venous resistance (PVR) greater than 3 Wood Units.[59] The prevalence of pulmonary hypertension in adult CHD has been reported between 4%-7%.[60,61] Prevalence of pulmonary hypertension changes with the type of adult CHD. Prevalence of pulmonary hypertension in adults with aortopulmonary window, atrioventricular septal defect, VSD and secundum ASD are 100%, 41%, 11% and 8%, respectively.[60] Data from Registry to EValuate Early And Long-term pulmonary arterial hypertension disease (REVEAL) management suggests that around 20% of all the patients with pulmonary hypertension have adult CHD.[62] Vasoconstriction, proliferation and obstructive remodelling, inflammation and thrombosis of the pulmonary vascular bed all appear to be involved in the development of pulmonary hypertension in CHD.[63,64] Along with erythrocytosis, pulmonary hypertension represents a physiologic adaptation to reduced oxygen delivery caused by admixture of deoxygenated blood as a consequence of right-to-left shunt. These patients have been recognised by the updated clinical classification of pulmonary hypertension as a separate entity in group 1 (Table 2).[65] An anatomic and pathophysiologic classification of congenital systemic-to-pulmonary shunts based on type and dimension of defects, direction of shunting and repair status had been introduced at the fourth World Symposium on Pulmonary Hypertension in 2008 (Table 3).[65] Overall, any patient with a lesion corrected too late or left untreated can develop pulmonary hypertension of variable severity and impaired outcome.

Eisenmenger syndrome is the most extreme manifestation of pulmonary hypertension seen in adult CHD like ASD, VSD and PDA. Eisenmenger syndrome is present in 58% of the patients with septal defect and pulmonary hypertension.[60] Dyspnoea is the most common feature followed by palpitation, oedema, haemoptysis, syncope and progressive cyanosis with the development of Eisenmenger physiology.[66] However, despite comparable morbidity, survival prospects for Eisenmenger patients are far superior to patients with pulmonary hypertension of other causes. Survival rates for this previously devastating condition before the advent of disease-targeting therapies are now reported at 75%, 70% and 55% at 30, 40 and 55 years of age, respectively.[67] It is largely unclear why this is so, but has been postulated that the integrity of the right ventricle is the major determinant of symptoms and survival rather than the degree of vascular injury.[68] Based on current data, practice guidelines recommend the use of targeted pulmonary hypertension therapy in Eisenmenger patients with New York Heart Association functional class III. While strongest evidence favours bosentan, the endothelin receptor antagonist, phosphodiesterase-5-inhibitors and prostanoids can also be considered as combination therapy in patients with insufficient improvement on single-drug therapy.[69-71]

Pulmonary Thromboembolism

Intrapulmonary thrombosis occurs in up to a third of adult patients with Eisenmenger physiology, particularly in Eisenmenger patients with large atrial septal defects.[72] Hypoxia in adults associated with Eisenmenger syndrome leads to secondary polycytharemia[73] and increase blood viscosity. This can induce *in-situ* thrombus formation in the pulmonary artery predisposing the patient to pulmonary infarction. Structural changes in the vessels, damage to the endothelium with activation of procoagulants, and impairment in the fibrinolytic system are other mechanisms that may contribute to thrombus formation in these individuals.[74] These patients presents with haemoptysis and pulmonary infarction.[75] Supportive treatments like hydration and iron supplements decreases hyperviscosity of the blood but ultimately may require anti-coagulants to prevent pulmonary thrombosis.[76]

Pulmonary Vascular Anomalies

Aside from the previously mentioned TAPVC seen with Scimitar syndrome,[53] pulmoniry stenosis is the major pulmonary valvular anomaly associated with CHD. Pulmoniry stenosis is present most commonly with LEOPARD syndrome,[43] Noonan's syndrome,[77] congenital rubella syndrome[78] and William's, syndrome.[34]

Pleural Disorders

Pneumothorax and pleural effusion are two important complications associated with adult CHD. These pleural disorders are either complication of connective tissue disorders or surgical procedures for CHD. These can be fatal, and hence, it is important to recognise them as early as possible.

Pneumothorax

Marfan's syndrome and EDS are the most common connective tissue disorders associated with adult CHD and pneumothorax.[79,80] Prevalence of spontaneous pneumothorax in Marfan's syndrome is

Table 2. Updated clinical classification of pulmonary arterial hypertension

Group 1: Pulmonary Arterial Hypertension

- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2
  - 1.2.2 ALK1, endoglin (with or without hereditary hemorrhagic telangiectasia)
  - 1.2.3 Unknown
- 1.3 Drug- and toxin-induced
- 1.4 Associated with
  - 1.4.1 Connective tissue diseases
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart diseases
  - 1.4.5 Schistosomiasis
  - 1.4.6 Chronic hemolytic anemia
- 1.5 Persistent pulmonary hypertension of the newborn
- 1′ Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis

Group 2: Pulmonary Hypertension Owing to Left Heart Disease

- 2.1 Systolic dysfunction
- 2.2 Diastolic dysfunction
- 2.3 Valvular disease

Group 3: Pulmonary Hypertension Owing to Lung Diseases and/or Hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental abnormalities

Group 4: Chronic Thromboembolic Pulmonary Hypertension

Group 5: Pulmonary Hypertension with Unclear Multifactorial Mechanisms

- 5.1 Hematologic disorders: myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis

Source: Reference 65

BMPE2=Bone morphogenetic protein receptor type 2; ALK1=Activin receptor like kinase type 1; HIV=Human immunodeficiency virus

Table 3. Classification of pulmonary hypertension in the setting of congenital heart disease

Presentation	Defect Size	Clinical Picture
Pulmonary hypertension with small defects	Atrial septal defect < 2cm Ventricular septal defect < 1cm	Similar to idiopathic pulmonary hypertension
Pulmonary hypertension with persistent left-to-right shunt	Moderate to large size defects	Left-to-right shunt No cyanosis at rest Mild to moderately elevated pulmonary arterial resistance
Eisenmenger syndrome	Large patent shunt lesions	Severely elevated pulmonary arterial resistance Bidirectional or right-to-left shunt, cyanosis, secondary erythrocytosis and multiorgan disease
Pulmonary hypertension after (late) corrective surgery	No or insignificant residual shunt lesion	Similar to pulmonary hypertension

Source: Reference 65

4.4% and frequently seen in males with apical blebs on the chest radiograph.[79] Recurrent primary pneumothorax is very common in these individuals, and hence, pleurodesis is recommended even after first episode of pneumothorax.[79]

Pneumothorax can develop as a consequence of adult CHD treatments like surgical interventions and implantable cardioversion defibrillators placement.[81,82] Pneumothorax is the second most common complication of surgical procedures for adult CHD, presenting in around 3.75% of total individuals treated with operative procedures.[81] Pneumothorax presents as early complication in 3.3% of total adult CHD individuals treated with cardiac defibrillators.[82]

Pleural Effusion

Heart failure is a common complication of adult CHD presents as bilateral pleural effusion. Congenitally corrected transposition of the great arteries, tetralogy of Fallot, Epstein's anomaly, subaortic stenosis, and aortic or mitral valve disease are associated with pleural effusion due to heart failure.[83] Heart failure associated with adult CHD should be treated with beta-adrenergic blockers and after-load reducers, and aldosterone antagonists and diuretics. Pleural effusion resolves with the improvement of heart failure.

Pleural effusion is present as a complication in around 0.85% adults surgically treated for CHD.[81] It may result from surgical procedure itself (non-specific effusion) or due to post-cardiac injury syndrome. Early post-operative complications like allergic response[84] or pulmonary embolism can also lead to pulmonary effusion. Pleural effusion my resolve spontaneously or occasionally require drainage.

Chylothorax is the complication of thoracic surgeries as these procedures may damage lymphatic duct. Surgery for CHD is the second most common cause of chylothorax among those surgical procedures.[85] Post-operative chylothorax can be managed conservatively with parenteral nutrition, use of medium chain triglyceride formula, low fat diet, octreotide or treated surgically with pleurodesis.[86]

Obesity Associated Pulmonary Manifestation

Prevalence of obesity/overweight among the patients with CHD is around 25%.[87] Individuals with CHD have normal to severely restricted physical activity which leads to obesity in these individuals.[88] Obesity can be associated with CHD as a part of complex disease syndromes like Down's syndrome, Turner's syndrome, Carpenter syndrome, Alstrom syndrome, Bardet-Biedl syndrome, and Isaacs' syndrome.[89-94]

Respiratory complications of obesity include asthma, chronic obstructive pulmonary disease (COPD), obstructive sleep apnoea (OSA) and obesity hypoventilation syndrome (OHS). Bronchial asthma increases by 10% and 7% in obese men and women, respectively.[93] Body mass index (BMI) is an important predictor of response to treatment in obese patients with asthma.[96,97] Increased BMI is associated with reduced mortality in patients with COPD.[98] On the other hand, weight gain has been associated with rapid decline in forced expiratory volume in COPD patients.[99] Dependent atelectasis, upper airway narrowing, restrictive effect of fat deposition on chest wall and abnormal respiratory drive are the causes of OSA and predispose to hypoventilation in individuals with obesity.[100]

Perioperative respiratory complications are very common among obese individuals. Obesity is associated with gastroesophageal reflux,[101] which increases the risk of aspiration pneumonia with anaesthesia. Large neck size is associated with difficult intubation and airway management.[102] Post-operative atelectasis is present in 45% of obese individuals.[103] Obese individuals also have a high risk of deep vein thrombosis and pulmonary embolism after surgery due to increased level of fibrinogen, factor VIII and von Willebrand factors.[104]

Pulmonary Functions Among Patients with Adult Congenital Heart Disease

Adults with CHD can have minimal to severe impairment of pulmonary function. Numerous factors are responsible for altered pulmonary function in patients with adult CHD. Chest wall abnormalities associated with adult CHD, like kyphoscoliosis, *pectus excavatum* and *pectus carinatum* causes restrictive pulmonary functions.[22,39] Pulmonary hypoplasia associated with pulmonic stenosis and SS is also an important factor for altered pulmonary function in adults with CHD.[54] Obesity associated with complex syndrome of CHD causes extrathoracic restrictive pattern.[89-94] Severity of derangement in pulmonary function is also determined by the nature of congenital heart lesion. Changes in pulmonary function are more marked in adults with cyanotic CHD as compared to adults with acyanotic CHD.[105] Pulmonary functions are also altered by diaphragmatic paralysis after corrective surgery for congenital heart lesions.[106]

Adults with CHD tend to have restrictive changes resulting in smaller volumes and flow rate, leading to a 20%-30% reduction in ventilatory capacity. Forced expiratory volume in the first second (FEV_1) and forced vital capacity (FVC) in these patients range from 71% to 84% of predicted values but FEV_1/FVC is preserved in almost all type of lesions. Total lung capacity in adults with CHD varies from 75%-81% of predicted values. Ratio of residual volume to tidal volume (RV/TLC) is slightly elevated above normal range in adults

with CHD but obstructive pattern of decreased FEV_1/FVC is not very common in these patients.[103] Adults with CHD have high oxygen requirement and altered pulmonary function, both of them increases work and cost of breathing in these individuals.

CONCLUSIONS

Prolonged survival in adult congenital heart disease is not only challenge for cardiologist but also for pulmonologist as pulmonary manifestations are very common in these individuals. We need more studies for better understanding of epidemiology and management of pulmonary diseases among adults with CHD. This review aims to emphasise the fact that that pulmonary manifestations should not be missed or neglected in adult patients with CHD as these play an important role in morbidity and mortality of these individuals.

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