University Heart Journal
Vol. 20, No. 1, January 2024

*REVIEW ARTICLE*

# Residua, Sequelae and Complications of Cardiac Defects in Adult Congenital Heart Disease after Repair at an Early Age

**MOHAMMAD ASHRAF UDDIN SULTAN, NAVEEN SHEIKH**

Department of Cardiology, Bangabandhu Sheikh Mujib Medical University Dhaka, Bangladesh.

**Address of Correspondence:** Dr. Mohammad Ashraf Uddin Sultan, Department of Cardiology, Bangabandhu Sheikh Mujib Medical University Dhaka, Bangladesh. E-mail: mausultan@gmail.com

**Abstracts:**

***A cardiac malformation, also known as a congenital heart defect, is a structural problem with the heart or blood vessels that is present at birth. These abnormalities occur when the heart or blood vessels don't form correctly in the womb. They are formed during the intra-uterine life and become visible between the 3rd and 8th week of pregnancy, when the heart is formed. Cardiac malformations include: Ventricular Septal defects, Transposition of the great vessels and Coarctation of the aorta. In most cases, the cause of cardiac malformations in unknown. Some know causes include: Maternal diabetes, Teratogens. It has been estimated that 45% of adults with congenital heart defects do not need routine follow-up in a specialized unit. For another 30%, the responsibility for care must be shared between the specialized unit and clinical cardiologist.***

***University Heart Journal 2024; 20(1): 31-35***
***DOI: https://doi.org/10.3329/uhj.v20i1.78482***

**Introduction:**

Cardiac malformations occur in approximately 1% of live births. Advances in surgery, interventional cardiology, and medical care have translated into increasing numbers of adult patients with congenital heart disease. These patients, even after intervention, have cardiac sequelae that require specialized care by cardiologists and cardiac surgeons with expertise in the management of congenital cardiac disease.[1]

Nowadays, it is estimated that 85% of the infants born with congenital heart disease (CHD) will survive to adulthood. The clinical profile and disease pattern of adults with CHD is changing. The prevalence of certain adult CHDs, such as tetralogy of Fallot, transposition of the great arteries or univentricular heart, is rising, but these conditions have practically become new diseases as a result of therapy. Most surviving patients present residua, sequelae, or complications, which can progress during adult life. These disorders can present electrophysiological disturbances, valvular disease, persistent shunts, myocardial dysfunction, pulmonary or systemic vascular disease, problems caused by prosthetic materials, infectious complications, thromboembolic events, or extravascular disorders involving multiple organs or systems.

In tetralogy of Fallot, the most striking problems that affect long-term prognosis are pulmonary valve regurgitation, right ventricle dysfunction, and atrial or ventricular arrhythmias.

The main problems appearing after physiological atrial repair of transposition of the great arteries are related to right ventricular function and atrial arrhythmias.

Surgical repair of univentricular heart using Fontan techniques should be considered a palliative procedure that does not modify the underlying structural disorder and exposes the postoperative patient to severe complications and problems.

The increase in the number of patients with CHD who will reach adulthood in the coming decades makes it necessary to carefully consider the new healthcare demands that are being generated, who should be responsible for them, and how and where solutions can be found.

**Abbreviations**

CHD: congenital heart disease.
ASD: atrial septal defect.
VSD: ventricular septal defect.
TF: tetralogy of Fallot.
TGV: transposition of great vessels.
ACHDU: adult congenital heart disease unit.

Arrhythmias and sudden death

In tetralogy of Fallot, the most striking problems that affect long-term prognosis are pulmonary valve regurgitation, right ventricle dysfunction, and atrial or ventricular arrhythmias.

Adults with operated TF experience frequent episodes of ventricular tachycardia or atrial tachyarrhythmia (flutter or fibrillation). These arrhythmias sometimes induce syncope or sudden death. A linear incidence of sudden

**Received:** 17 November, 2023 **Accepted:** 15 December, 2023

death of 0.4% annually during the first 25 years after the intervention has been estimated, and the incidence grows exponentially from the first 25 years on.[1] The arrhythmia substrate may be related to surgical scars, although the dilation of the right ventricle and atrium induced by the pulmonary and tricuspid insufficiency acts as a trigger factor.[2] It has recently been confirmed that the patients with the greatest risk of atrial and ventricular arrhythmias, including sudden death, are those that have severe pulmonary and/or tricuspid insufficiency and develop a marked cardiomegaly, progressive widening of the QRS complex, and an increase in the dispersion of the QT interval.[3,4] A cardiothoracic index >60%, QRS duration >180 ms and QT dispersion >60 ms are markers of risk that are easy to obtain at the bedside of the patient.[5]

**Residual shunts**

Residual shunts are not infrequent. Residual VSD after surgical repair is detected frequently on postoperative echocardiogram. Residual VSD with a mean diameter of 3 mm, greater may be less likely to close spontaneously after three years .The most frequent location is the interventricular patch, although some patients may have an extracardiac shunt related with surgical fistulas or unligated aortopulmonary collateral vessels. Percutaneous VSD closure is a well-established procedure with a low severe adverse event rate. There are two risk factors for residual shunting: VSDs after surgical closure and use of Oversized Membranous Amplatzer occluder may cause complete heart block .

Atrial Septal Defect (ASD) is one of the most common congenital cardiac defect. Even though surgical repair of ASD is the current method of choice but percutaneous device closure is rapidly gaining popularity as it is less invasive. Dislodgment and embolization of the device may occur requiring urgent surgical retrieval. Some patients may have a permeable foramen ovale or an ASD not closed in the correction. This has been cause of paradoxical embolism in some cases. Most residual interatrial or aortopulmonary shunts can be treated with percutaneous procedures.

Residual shunts in post ligation PDA can occur in approximately 6% of cases. Even though spontaneous closure of silent PDA might occur within 3-6 months, 7% of patients will require a second procedure to close the shunt (6). Patients with a small and hemodynamically insignificant PDA often remain asymptomatic and may never develop symptom.[7]

**Reintervention in adulthood**

Very recent findings suggest that surgical reintervention to correct pulmonary insufficiency with a valvular prosthesis and tricuspid insufficiency by annuloplasty decreases the incidence of atrial and ventricular arrhythmias, especially if surgical repair is accompanied by ablation of the reentry circuit.[8] In most cases, the dilation and systolic dysfunction of the right ventricle do not remit after surgery.

**Dysfunction of the systemic right ventricle**

Patients with TGA have a special anatomic situation in which the right ventricle is connected to the aorta and must bear systemic circulation. The use of the Mustard and Senning techniques for atrial physiological repair means that many patients have a right ventricle that is surgically connected to the aorta. In the postoperative follow-up, a large number of patients present systolic dysfunction of the right ventricle at rest or after physical exercise.[9,10,11] As adults life, most of them are clinically well, but have a reduced capacity for effort.[12] Some develop chronic heart failure .The cause of postoperative right ventricular dysfunction is multifactorial.[13]

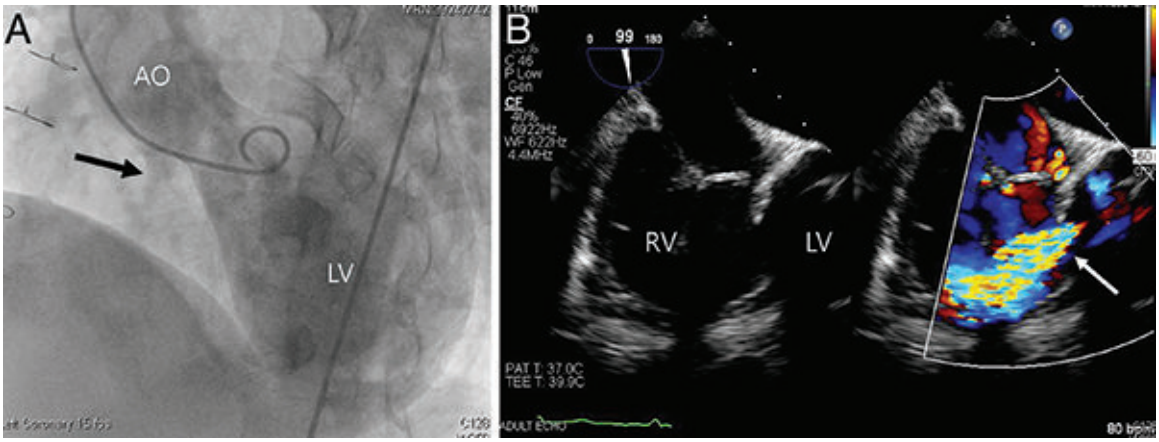


**Figure 1:** *Device closure of residual postopertive VSD with Coccoon VSD occlude device. a LV angiography showed residual VSD (black arrow). b transesophageal echocardiography and color Doppler revealed residual VSD (white arrow) with significant amount.*

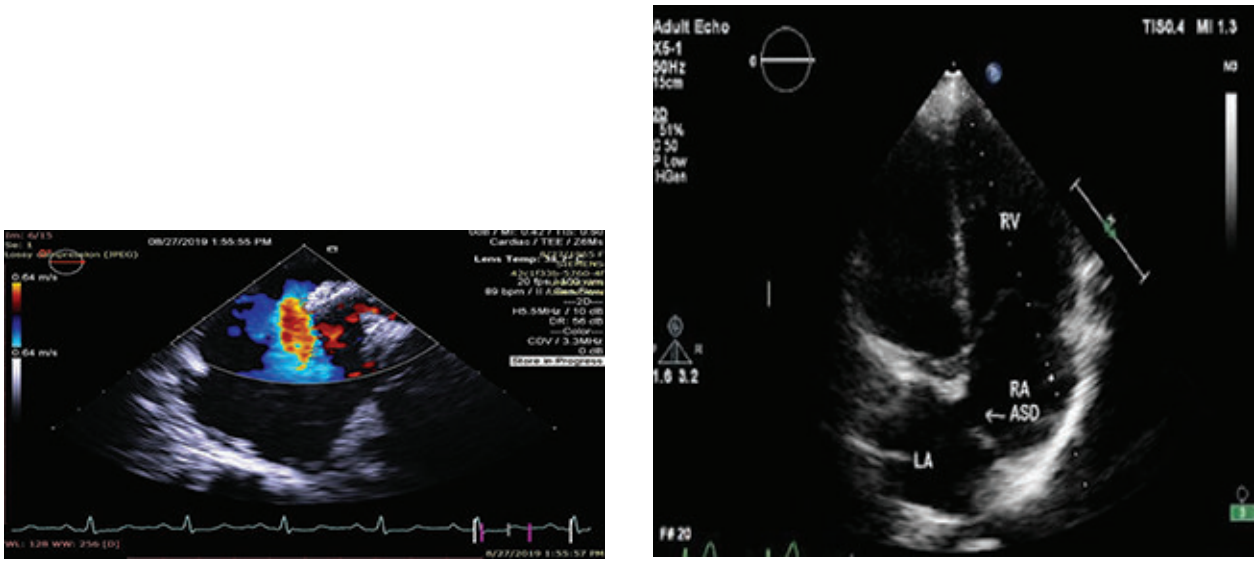


**Figure 2:** *(a) Transoesophageal echocardiogram with colour doppler showing presence of left to right shunting across the atrial septal defect. (b) Echo view of residual atrial septal defect*

**Cardiac rhythm disorders**

The most frequent rhythm disorders atrial switch are symptomatic sinus bradycardia, slow nodal rhythms, and paroxysmal or sustained supraventricular tachycardia, principally atrial flutter. The cause lies in the lesion of the sinus node during surgery and the atrial scars that interrupt the preferred internodal conduction pathways, which form the substrate for reentry circuits.[14]

The current indications for a permanent pacemaker in patients with TGA are complete AV block, sustained heart rates of less than 30 beats/min, documented Stokes-Adams episodes, severe ventricular systolic dysfunction with bradycardia, and the need to begin treatment with antiarrhythmic drugs that depress sinus function even more.

**Valvular dysfunction**

Insufficiency of the tricuspid valve, which is located on the systemic ventricle, can appear after surgical atrial correction. This complication is more prevalent in patients with a large VSD that has been closed with a septal patch.[15] Some patients present stenosis of the left ventricular outflow tract of subvalvular location.

**Arterial correction**

Arterial correction with the Jatene technique is replacing atrial correction in most hospital centers. The most frequent complication is pulmonary supravalvular stenosis[16] followed by the less frequent aortic supravalvular stenosis. This dilation can induce annulectasia However, a relatively high incidence of stenosis or occlusion of main coronary arteries has been detected even in patients who survive the intervention with no apparent perioperative complications[17,18]

**UNIVENTRICULAR HEART**

The term univentricular heart (UH) defines complex congenital heart disease that lacks a pulmonic ventricular chamber either in the original anatomy or the final palliation. The prevalence of patients with this type of physiology continues to increase due to improved surgical palliative procedures.

**From Glenn to Fontan**

In 1958, Glenn established the therapeutic basis for eliminating the right side of the heart from the pulmonary circuit as an alternative to systemic-pulmonary fistulas(19). Glenn´s original intervention consisted of end-to-end anastomosis between the superior vena cava and right pulmonary artery.

**Obstruction and thrombosis**

The atrial or cavopulmonary connections may become obstructed by stenosis or kinking of the pulmonary arteries, retraction of suture lines, intrinsic degeneration of valvular prostheses, neointimal proliferation of conduits, calcification and the rigidity of prosthetic materials, or intraluminal thrombosis. The high incidence of thrombosis found in some series suggests that most adult patients should receive permanent anticoagulant treatment if no specific contraindications are present(20). Another long-term complication is the obstruction of the left ventricular outflow tract.

**Persistent shunts and valvular insufficiency**

After Fontan surgery, many patients have persistent shunts. The insufficiency can appear in very late phases of evolution and may have a progressive nature.[21]

Atrial arrhythmias and congestive heart failure

Atrial arrhythmias, especially atrial flutter and tachycardia due to macroreentry. The prevalence of atrial arrhythmias in the follow-up of adults with Fontan physiology varies from 40% to 60% in different studies.[22,23]

**Valvular dysfunction**

Insufficiency of the tricuspid valve is more prevalent in patients with a large VSD that has been closed with a septal patch.[24]

**Arterial correction**

Arterial correction with the Jatene technique is replacing atrial correction in most hospital centers.[25] The most frequent complication is pulmonary supravalvular stenosis[26,27] followed by the less frequent aortic supravalvular stenosis.

Single-Ventricle Heart

These are generally serious and complex CHDs with scant natural survival to adulthood and cardiologists for adults are not very familiar with them. Cardiologists who treat adults must approach them as if they were a homogeneous disease.

**From Glenn to Fontan**

In 1958, Glenn established the therapeutic basis for eliminating the right side of the heart from the pulmonary circuit as an alternative to systemic-pulmonary fistulas.[28] Glenn´s original intervention consisted of end-to-end anastomosis between the superior vena cava and right pulmonary artery.

**Obstruction and thrombosis**

The main complications of long-term Fontan are atrial or cavopulmonary connections may become obstructed by stenosis or kinking of the pulmonary arteries, retraction of suture lines, intrinsic degeneration of valvular prostheses, neointimal proliferation of conduits, calcification and the rigidity of prosthetic materials, or intraluminal thrombosis.[29]

**Persistent shunts and valvular insufficiency**

After Fontan surgery, many patients have persistent shunts that can be produced by dehiscence of patches , aortopulmonary anastomoses, or the development of pulmonary arteriovenous fistulas.

**Protein-losing enteropathy**

One of the worst long-term consequences of the Fontan operation is protein-losing enteropathy, which produces refractory ascites, generalized edema, and massive pleural effusion.[30]

**Revision of the Fontan operation**

In patients with symptomatic atrial arrhythmias or severe protein-losing enteropathy that does not respond to medical treatment, revision of the Fontan intervention in adult life can be necessary.

**Needs of Adults with Congenital Heart Disease**

The large increment in the number of patients with CHD who will reach adulthood in the coming decades demands careful consideration of the new needs for care that are being generated, who will be responsible for providing care, and where it will be carried out. The physiology of many previously repaired or palliated CHDs can be difficult to understand for cardiologists .

In every hospital, ACHDU should be formed by at least one cardiology specialist and an expert in CHDs, heart surgeons expert in pediatric and adult problems, as well as anesthetists with training and experience in both fields. Invasive (catheterization, electrophysiology) and non-invasive studies (echocardiography, stress techniques, radionuclide medicine, magnetic resonance imaging) must be performed preferentially in adult units by specialists who are experts in CHDs. The ACHDU should also have access to a clinic for high risk pregnancies, a cardiac rehabilitation unit with special knowledge of congenital problems. A fluid relation with the areas of clinical genetics (genetic counseling), hematology (blood dyscrasias), neurology (neurological syndromes), traumatology (skeletal malformations), and other specialized areas is important.

It has been estimated that 45% of adults with CHD do not need routine follow-up in a specialized unit. For another 30%, the responsibility for care must be shared between the specialized unit and clinical cardiologist.[31] Ideally, each adult patient with CHD would have to be examined at least once in an ACHDU and returned to the community if a highly specialized follow-up is not necessary.

**References:**

1. Nollert G, Fischlein T, Bouterwek S, Bohmer C, Klinner W, Reichart B. Long-term survival in patients with repair of tetralogy of Fallot: 36-year follow-up of 490 survivors of the first year after surgical repair. J Am Coll Cardiol, 30 (1997), 1374-83.
2. Zahka KG, Horneffer PJ, Rowe SA, Neill CA, Manolio TA, Kidd L, et al. Long-term valvular function after total repair of tetralogy

of Fallot: relation to ventricular arrhythmias. Circulation, 78 (1988), III14-9

3. Gatzoulis MA, Till JA, Somerville J, Redington AN. Mechanoelectrical interaction in tetralogy of Fallot: QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death. Circulation, 92 (1995), 231-7
4. Gatzoulis MA, Till JA, Redington AN. Depolarization-repolarization inhomogeneity after repair of tetralogy of Fallot: the substrate for malignant ventricular tachycardia Circulation, 95 (1997), 401-4
5. The adult with tetralogy of Fallot. Armonk: Futura Publishing Company, Inc., 2001.
6. Follow-up after surgical closure of congenital ventricular septal defect European Journal of Cardio-Thoracic Surgery, Volume Issue 4, October 2003, Pages 511–15, https://doi.org/10.1016/S1010-7940(03)00430-5
7. Liang C Di, Ko SF, Huang CF, Chien SJ, Lin YJ, Lin IC *et al*. Immediate echocardiographic surveillance after transcatheter closure of a patent ductus arteriosus: a feasible method to assess residual Shunt. Pediatr Neonatol. 2010 Feb;51-52
8. Therrien J, Siu SC, Harris L, Dore A, Niwa K, Janousek J, et al..Impact of pulmonary valve replacement on arrhythmia propensity late after repair of tetralogy of Fallot..Circulation, 103 (2001), 2489-94
9. Adults with congenital heart disease and previous intervention Sabrina D Phillips, Crystal R Bonnichsen, Christopher J McLeod, Naser M Ammash, Harold M Burkhart, Heidi M Connolly DOI: 10.1016/j.cpcardiol.2013.05.002
10. Hagler DG, Ritter DG, Mair DD, David GD, McGoon DC.Clinical, angiographic, and hemodynamic assessment of late results after Mustard's operation. Circulation, 57 (1978), 1214-20
11. Kato H, Nakono S, Motsada H, Hirose H, Shimazaki Y, Kawashima Y.Right ventricular myocardial function after atrial switch operation for transposition of the great arteries.Am J Cardiol, 63 (1989), 226-30
12. Reybrouck T, Dumoulin M, Van der Hauwaert LG. Cardiorespiratory exercise testing after venous switch operation in children with complete transposition of the great arteries. Am J Cardiol, 61 (1988), 861-5
13. Hurwitz RA, Caldwell RL, Girod DA, Mahony L, Brown J, King H.Ventricular function in transposition of the great arteries: evaluation by radionuclide angiocardiography.Am Heart J, 110 (1985), 600-5
14. Gillete PC, Kugler JD, Garson Aj.r, Gutgesell HP, Duff DF, McNamara DG.Mechanisms of cardiac arrhythmias after the Mustard operation for transposition of the great arteries.Am J Cardiol, 45 (1980), 1225-30
15. Fricker FJ, Griffith BP, Hardesty RL, Trento A, Gold LM, Schmeltz K, et al. Experience with heart transplantation in children. Pediatrics 1987;79:138-46.
16. Van Praagh R. Diagnosis of complex congenital heart disease: morphologic-anatomic method and terminology. Cardiovasc Intervent Radiol 1984;7:115-20.
17. Anderson RH, Becker AE, Freedom RM, Macartney FJ, QueroJiménez M, Shinebourne EA, et al. Sequential segmental analysis of congenital heart disease. Pediatr Cardiol 1984;5: 281-7.
18. Tynan MJ, Becker AE, Macartney FJ, Quero-Jiménez M,Shinebourne EA, Anderson RH. Nomenclature and classification of congenital heart disease. Br Heart J 1979;41: 544-53.
19. Bargeron LM, Elliot LP, Soto B, Bream PR, Curry GC. Axial Cineangiography in congenital heart disease. I: Concept,technical and anatomic considerations. Circulation 1977;56:1075-83.
20. Elliot LP, Bargeron LM, Bream PR, Soto B, Curry GC. Axial Cineangiography in congenital heart disease. II: Specific lesions. Circulation 1977;56:1084-93.
21. Gutgesell HP, Huhta JC, Latson LA, Huffines D, McNamara DG. Accuracy of two-dimensional echocardiography in the diagnosis of congenital heart disease. Am J Cardiol 1985;55:514-8.
22. Treves ST, Newberger J, Hurwitz R. Radionuclide angiocardiography in children. J Am Coll Cardiol 1985;5:120-122
23. Irsik RD, White RD, Robitaille PM. Cardiac magnetic resonance imaging. En: Moss-Adams, editor. Heart disease in infants, children, and adolescents. Baltimore: Williams & Wilkins, 1995; 206-23.
24. Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy: a palliative approach to complete transposition of the great arteries. JAMA 1966;196:991-2.
25. Rashkind WJ. Transcatheter treatment of congenital heart disease. Circulation 1983;67:711-6.
26. Mullins CE, O'Laughlin MP. Therapeutic cardiac catheterization. En: Moss-Adams, editor. Heart disease in infants, children, and adolescents. Baltimore: Williams & Wilkins, 1995; 439-52.
27. Romero R, Pilu G, Jeanty P, Ghidini A, Hobbins JC. Prenatal diagnosis of congenital anomalies. East Norwalk: Appleton & Lange, 1988
28. Moller JH, Anderson RC. 1,000 consecutive children with a cardiac malformation with 26 to 37 years follow-up. Am J Cardiol 1992;70:661-7.
29. O'Fallon WM, Crowson CS, Rings LJ, Weidman WH, Driscoll DJ, Gersony WM, et al. Second natural history study of congenital heart defects. Materials and methods. Circulation 1993; 87(Suppl):I4-I15.
30. Hayes CJ, Gersony WM, Driscoll DJ, Keane JF, Kidd L,O'Fallon WM, et al. Second natural history study of congenital heart defects: results of treatment of patients with pulmonary valvar stenosis. Circulation 1993;87(Suppl):I28-I37.