

Congenital Heart Disease: A Concise Review on Various Testing Methods

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Abstract - Congenital Heart Disease (CHD) is a medical terminology applicable for the deformity present in a child's heart or if the blood vessels fail to function during its birth. The deformity might affect the child's blood flow through the heart. Even in today's times of modern technology, it has been noticed that it is onerous to detect congenital heart disease significantly. It is a rarely diagnosed and its prevalence is for 0.5% - 1% of the total population. It is crucial that accurate methods are practiced figuring out the same. This paper focuses on various testing methods to detect Valvular heart disease, Brugada Syndrome (BrS), Ventricular Arrhythmia (VT), Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA), Wolff-Parkinson-White Syndrome. The results obtained by comparing these diseases using magnetic resonance imaging (MRI), Computed Tomography (CT) scan, Electrocardiogram (ECG) and Echocardiogram are analyzed in this paper. Analysis suggests that these diseases are rarely survived without treatment and are required to surpass the early years of life for successful treatment.

Key Words: Haemodynamic, Congenital Heart Disease, Ventricular Arrhythmia, ALCAPA, Wolff-Parkinson-White Syndrome, Valvular heart disease.

1. INTRODUCTION

Heart Disease

One of the most sensitive organ of the human body is nothing but the heart. Malfunctioning of the heart or any minor harm to it can prove fatal. A group of diseases that affect the heart and associated blood vessels is termed heart diseases. The risk of certain heart disease may be increased by smoking, high blood pressure, unhealthy heart, lack of exercise, obesity etc. Similarly, many birth abnormalities cause aberrant physiology, which can lead to physical, developmental, or intellectual disabilities. The phenotype that results might range from minor impairment to severe incompatibility with life. The foetus is incompatible with life in the majority of cases, and it is spontaneously aborted.

Birth abnormalities can be classified as structural, altering the body's "form," or functional, impacting

the "function" of an organ or body system, in this case, the heart and its circulation. Congenital Heart Disease (CHD) is a non-specific medical term for a range of defects present at the time of birth that affect the normal physiology of the heart and associated cardiovascular system. CHD is a structural defect of the heart that may change or impede normal blood flow. CHD occurs when the heart or the blood vessels near the heart fail to develop properly before birth.

Classification of CHD

There are many diverse types of CHDs. Some simpler forms of CHDs may have no symptoms and require no treatment, while more serious and complex defects may have severe or life-threatening symptoms that require medical or surgical intervention. The anatomy of congenital cardiac malformations, either pre- or post-operative, can vary from simple to complex to critical. Many CHD types require (percutaneous or surgical) intervention, either corrective or palliative. With the improvement of various surgical and interventional techniques, the patients' survival rate has increased dramatically.

A class of CHD based on rarely monitored includes Brugada syndrome, Ventricular septal defect, Valvular heart disease, Ventricular Arrhythmia, ALCAPA Syndrome, Wolff Parkinson syndrome and many more.

1.3.1 Brugada syndrome:

An inherited cardiac disease that increases the risk of ventricular tachyarrhythmias and sudden cardiac death in otherwise healthy young adults. Brugada syndrome (BrS) is a genetic disorder characterized by abnormal electrical activity in the heart. It raises the risk of irregular heartbeats and sudden cardiac death. Brugada syndrome patients typically have dangerous arrhythmias like ventricular fibrillation or polymorphic ventricular tachycardia, but they're also more likely to have rapid heart rates due to less

dangerous arrhythmias like AV nodal reentrant tachycardia and abnormally slow heart rhythms like sinus node dysfunction. There are several mechanisms by which the genetic mutations causing this condition might produce these arrhythmias.

1.3.2 Valvular heart disease:

When any valve in the heart is damaged or diseased it is termed as Valvular heart disease. These problems are primarily caused by ageing, although they can also be caused by congenital (inborn) anomalies, particular disease, or physiologic processes such as rheumatic heart disease and pregnancy. The valves are anatomically part of the dense connective tissue of the heart known as the cardiac skeleton and are responsible for regulating blood flow through the heart and major arteries. Valve failure or malfunction can lead to a reduction in heart function, albeit the specific effects vary depending on the kind and degree of the valvular disease.

1.3.3 Ventricular Arrhythmia:

State of the body when the lower heart chambers called ventricles originate abnormal heartbeats is termed as ventricular arrhythmia. This condition may also be called V-tach or VT. At rest, a healthy heart beats roughly 60 to 100 times per minute. The heart beats faster than normal in ventricular tachycardia, usually 100 or more beats per minute. The irregular heartbeats prevent the heart chambers from filling adequately with blood. As a result, the heart may be unable to pump enough blood to the body and lungs.

1.3.4 ALCAPA Syndrome:

Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA), also known as Bland-White-Garland syndrome or White-Garland syndrome, is a rare congenital defect in which the Left Coronary Artery (LCA) branches from the pulmonary artery rather than the aortic sinus. Following birth, the pressure in other coronary arteries (particularly the RCA) will exceed the pressure in the LCA, increasing collateral circulation. As a result, blood can flow from the RCA to the LCA and then to the pulmonary artery, forming a left-to-right shunt.

1.3.5 Wolff-Parkinson White Syndrome:

Wolff-Parkinson-White syndrome (WPWS) is a condition caused by a specific sort of malfunction with the heart's electrical system, which causes

symptoms. An auxiliary electrical conduction route between the atria and the ventricles is the fundamental mechanism.

1.1 Testing Methods

A CHD is often detected during a pregnancy ultrasound. If a doctor hears an abnormal heartbeat, for instance, they may further investigate the issue by performing certain tests. These may include an echocardiogram, electrocardiogram, a chest X-ray, or an MRI scan. Blood tests are rarely used to detect CHD's. When sound waves are used to produce images of the heart the method is termed as echocardiography while when electrical activity is used to check the heart's rhythm, it is called electrocardiography. The method where X-rays are used to detect a disease is termed as CT scan while MRI uses strong magnetic fields and radio waves.

1.4.1 ECHOCARDIOGRAPHY

Echocardiography is a group of interrelated applications of ultrasound including two-dimensional anatomical imaging, M-mode echocardiography, Doppler techniques, and contrast echocardiography. Echocardiography uses sound in the frequency range of 1 to 10 MHz. Typically, when imaging adult patients, frequencies range from 2 to 5 MHz. For pediatric and some specialized adult applications higher frequencies of 7.5 and 10 MHz may also be used.

Principle of working:

Echocardiography is a type of medical imaging of the heart using standard ultrasound or Doppler ultrasound. The actual clinical echocardiographic examination consists of simultaneous and integrated two-dimensional and Doppler evaluation, supplemented by other specific and goal-oriented imaging modalities.

The underlying premise of cardiac ultrasonography is that the speed of sound through tissue is equal to that in water (1450 m/sec). Ultrasound scanners generate a series of ultrasound bursts at a set frequency. The energy from the ultrasound is then reflected back to the transducer by cardiac and other structures. The distance between a reflective object and a transducer can be calculated by calculating the time required for round trip transit.

M-mode echocardiography was the first type of clinical cardiac ultrasonography. M-mode echocardiography relies on interrogation along a

single line of ultrasound either emitted from an independent transducer or along a cursor within a two-dimensional image. The M-mode echocardiogram only provides information with respect to the distance of each object from the transducer and provides no information in the lateral dimension.

A two-dimensional echocardiogram is created when a fan-shaped array, consisting of multiple interrogation lines, is emitted typically over a 90-degree sector from the transducer and analyzed. There are two methods by which the fan-shaped sector of ultrasound beams can be created. Most widely used is a phased-array system in which a linear array of ultrasound crystals is electronically steered through an imaging arc. The second method relies on high-speed mechanical rotation of one or usually more ultrasound crystals to mechanically steer the ultrasound beam through the predefined arc.

Doppler ultrasonography relies on analysis of a shift in the frequency of the ultrasound beam due to interaction with moving targets. The Doppler shift data can be displayed as a velocity profile or a color flow image.

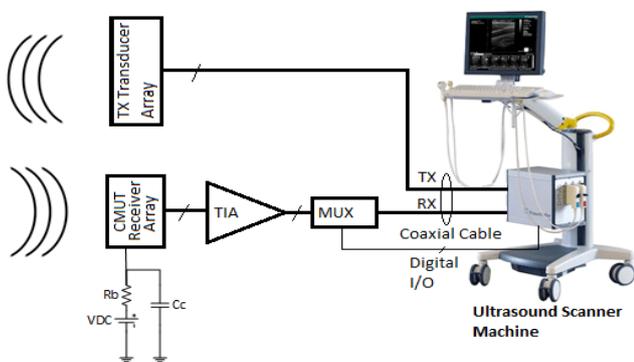


Fig -1: Block diagram of Ultrasound Scanner machine.

1.4.2 ELECTROCARDIOGRAPHY

The electrocardiogram (ECG), as used today, is the product of a series of technological and physiological advances pioneered over the past two centuries. Early demonstrations of the heart's electrical activity, for example, by Marchand and others, were closely followed by direct recordings of cardiac potentials by Waller in 1887, which has become a widely used and invaluable clinical tool for the detection and diagnosis of a wide range of cardiac conditions, as well as a technique that has contributed to a better

understanding of the heart. In addition, the ECG is critical in the treatment of major metabolic abnormalities. Moreover, it has remained the most direct method for assessing abnormalities of cardiac rhythm. Use of the ECG for any of these clinically important purposes is the final outcome of a complex series of physiological and technological processes.

Principle of working:

First, an extracellular cardiac electrical field is generated by ion fluxes across cell membranes and between adjacent cells. These ion currents are synchronised by cardiac activation and recovery sequences to produce a varying cardiac electrical field in and around the heart during the cardiac cycle. This electrical field passes through numerous other structures, including the lungs, blood, and skeletal muscle, before reaching the body surface. These structures are known as transmission factors which differ in their electrical properties and perturb the cardiac electrical field as it passes through them. Electrodes placed in specific locations on the extremities and torso and configured to produce leads detect the potential reaching the skin. The outputs of these leads are amplified, filtered, and displayed by a variety of electronic devices to construct an ECG recording. Finally, diagnostic criteria are applied to these recordings to produce an interpretation. The criteria have statistical characteristics that determine the clinical utility of the findings. Each of these steps influences the final product -the clinical ECG. [3]

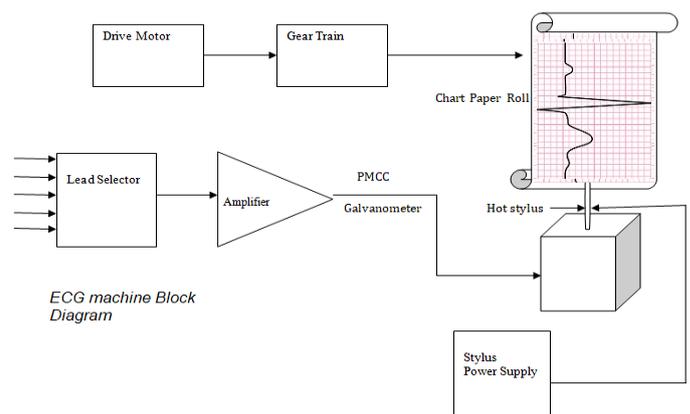


Fig -2: Block diagram of ECG machine.

1.4.3 CT-SCAN

Computed Tomography of the heart usually requires modification of the standard CT techniques used for investigating other parts of the body. Newer spiral and multiple array CT scanners with exposure times

of less than 1 second are usually adequate for some purposes, such as evaluating thoracic aortic disease, pericardial disease, para-cardiac and intracardiac tumours, and the patency of coronary arterial bypass grafts. Continuously rotating (spiral) CT scanners have an exposure time of 1 second or less for each image with no interscan delay between images at sequential anatomical levels, producing images of the entire heart in 12 to 20 seconds. Multiple-array CT scanners can acquire images of the entire heart and proximal aorta in several seconds. Although adequate anatomical depiction of cardiovascular anatomy is attained with spiral and multidetector CT scanners, scans corresponding to precise phases of the cardiac cycle cannot be obtained. For the assessment of cardiac dimensions and function in addition to morphology, millisecond CT scanners are required

Principle of working:

Computed tomography can detect not only the inner endocardial wall but also the epicardial surface. Wall thickness and myocardial mass were calculated precisely. There is a strong correlation between CT measurements and postmortem anatomical measurements of wall thickness and mass. It has also been used to estimate right ventricular mass by measuring the free wall mass.

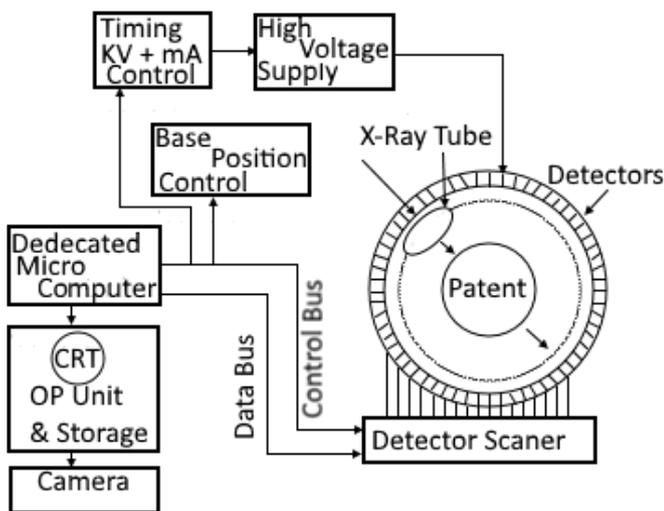


Fig -3: Block diagram of CT scan machine.

A series of tomograms in a short-axis plane acquired during multiple phases of the cardiac cycle permits measurement of area ejection fraction and wall thickening at various levels of the left ventricle, extending from the base to the apex. In normal

human subjects, a variation in both regional ejection fraction and extent of wall thickening has been defined, a gradient in both area ejection fraction and extent of wall thickening increases progressively from basal to apical layers. In global and regional myocardial abnormalities, cine CT has revealed dysfunction of wall thickening and wall motion.

1.4.4 MRI

Magnetic resonance imaging (MRI) has several important characteristics that make it an excellent tool for cardiovascular diagnosis. Because of the lack of signal from moving blood with the spin-echo MRI technique or the vivid signal from blood with the gradient-echo (cine MRI) approach, there is a significant natural contrast between the blood pool and the cardiovascular structures. When the spin-echo technique is used, blood appears black on images; thus, internal structures of the heart could be visualized within the signal void of the cardiac chambers. With the use of the gradient-echo technique, the blood pool appears white and has substantially higher signal than the myocardium, again providing a good edge definition of the endocardial margin. As a result, no contrast medium is required for blood pool discrimination, making MRI a completely noninvasive imaging technique.

A broad range of soft tissue contrast allows for the characterization of myocardial tissue. The density of protons (hydrogen nuclei), magnetic relaxation times of the protons, and magnetic susceptibility effects all influence tissue contrast. The use of MR contrast media improves the characterization of myocardial tissue. Imaging can be performed in any plane, including those parallel and perpendicular to the ventricle's major axis. With velocity encoded MRI, blood flow can be measured in any cardiac chamber or blood vessel.

Principle of working:

Atomic nuclei with a net charge have a magnetic moment. A net charge exists when a nucleus contains unpaired (an odd number) protons, neutrons, or both. The hydrogen nucleus contains only a proton; it is positively charged and has a strong magnetic moment. The magnetic properties of nuclei are expressed when they are placed in an external magnetic field. When protons or other nuclei with magnetic moment lie within a magnetic field and are then exposed to electromagnetic radiation (RF waves), energy is absorbed and subsequently emitted. This absorption and release of energy

causes resonance-nuclear MR. The RF necessary to induce resonance has to be proportional to the local magnetic field (HL) and a constant (magnetogyric ratio [g]) related to the specific nucleus involved. The relationship between frequency (f) and magnetic field is expressed by the following equation:

$$f = gHL / 2\pi$$

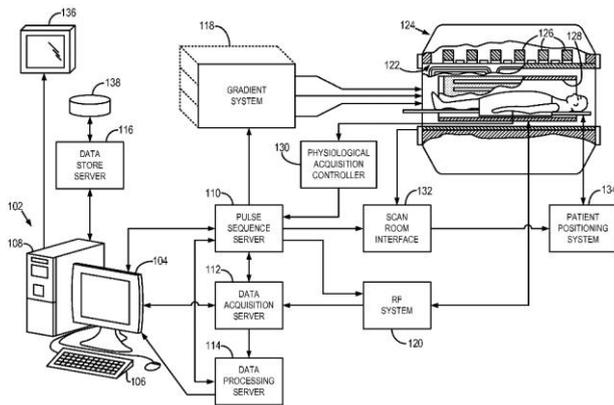


Fig -4: Block diagram of MRI machine.

When nuclei at equilibrium in a magnetic field are irradiated at the resonant frequency, they attain a higher energy state. When they return to equilibrium, they emit energy at the same frequency if the magnetic field remains constant. If the magnetic field changes between the time of excitation and emission, then the emission occurs at a frequency corresponding to the new field strength.

2. MOTIVATION:

The greatest cause of death today is cardiovascular disease (CVD), which encompasses a wide spectrum of cardiovascular system disorders that impact heart function. The early detection of CVD is thought to minimise the disease's high death and morbidity rates. As a result, a collection of comprehensive clinical cardiovascular assessment tools has been established to compute cardiovascular haemodynamics in order to provide useful insights to clinicians in recognising indications that contribute to CVD and assisting in CVD diagnosis.. Cardiovascular disease is the most commonly reported cause of mortality, and it contributes to approximately 30% of deaths worldwide. Cardiovascular disease (CVD) accounted for fewer than 10% of all deaths globally at the turn of the twentieth century. CVD accounted for approximately half of all fatalities in the industrialised world and 25% in the developing world by the end of the study.

By 2020, cardiovascular disease (CVD) will account for 25 million deaths per year, and coronary heart disease (CHD) will overtake infectious disease as the leading cause of death and disability worldwide. The most prevalent birth abnormalities are congenital heart disorders (CHDs), which account for roughly one-third of all congenital birth defects.

Despite the number of studies that have been carried out on blood flow in LV, cardiac modelling and blood flow across LV is still not understood well. Various approaches such as echocardiography, magnetic resonance imaging etc. have been adopted to visualize flow patterns through LV to understand fluid dynamics in LV of the heart. CMR (Cardiovascular Magnetic Resonance) is emerging as an alternative tool to further ease the visualization of patient-specific LV flow, as it can provide more detailed flow structure with much higher resolutions.

3. Literature Survey

3.1 Brugada syndrome

Brugada syndrome is a hereditary condition that can result in a dangerously erratic heartbeat. Due to this condition, your heart's lower chambers (ventricles) beat rapidly and erratically. This inhibits blood from flowing freely throughout your body. This is dangerous since it can cause fainting or even prove fatal, especially while sleeping or resting. Since victims with the disease frequently die while sleeping, it's been known as sudden, unexplained nocturnal death syndrome. Brugada syndrome is an uncommon condition. It affects around 5 out of every 10,000 people in the world. Adulthood is when most symptoms appear. However, the illness can strike at any age, including childhood. It is possible to have Brugada syndrome that is dormant and does not cause any symptoms. However, several medications, such as antidepressants and antipsychotics, as well as illegal narcotics, fever-causing diseases, and electrolyte imbalances, can trigger the syndrome.

3.1.1 Brugada- MRI

On cine images, there was regional contractility (hypokinesis, akinesis, dyskinesia) of the left ventricle (LV), as well as fatty infiltration of both ventricles on turbo spin echo (TSE) images. The regional contractility of the right ventricle (RV) was quantified by measuring radial fractional shortening in each of eight segments in which the RV-free wall was separated (three basal, three mid-ventricular, and two apical). At the end of the diastole, around 1 cm below the AV valve plane, only a few mono-

dimensional measurements of both the LV and RV were recorded. [4]

3.1.2 Brugada- CT scan

The right ventricle's morphological defects were revealed by an electron beam CT scan. The appearance of premature ventricular contractions recorded exclusively in the acute phase, which may precipitate ventricular fibrillation, was linked to the areas of morphological abnormalities revealed by electron beam CT in individuals with the Brugada syndrome. In cases of the Brugada syndrome, these morphological abnormalities may be linked to arrhythmogenic substrates. [5]

3.1.3 Brugada- ECG

Mechanical dispersion, which was calculated in all patients as a measure of contraction heterogeneity, was defined as the standard deviation of the time interval from the onset of the QRS-complex until the peak myocardial shortening in the 16-left ventricle, 3 right ventricle free wall, and 6 right ventricle segments.

In the BrS, the conventional 12-lead ECG is critical for diagnosis and, more than likely, for regulating the prognosis (i.e., the amount of arrhythmic risk). The BrS affects up to 40% of patients who have a normal or nondiagnostic resting ECG. Using currently available ECG-based approaches, the diagnosis of BrS is rather simple. In BrS, successful ECG-based risk classification was achieved. [6]

3.2 Valvular heart disease -

The heart contains four valves through which the blood keeps flowing in the proper direction. One or more of the valves may not open or close properly in some cases. This can disrupt the flow of blood from your heart to your body. Birth defects in heart valves are possible (congenital). It can also occur in adults as a result of a variety of causes and conditions, including infections and other heart conditions.

Problems with heart valves may include:

Regurgitation. The valve flaps do not close properly in this condition, allowing blood to leak backward into your heart. This is most commonly caused by valve flaps bulging back, a condition known as prolapse.

Stenosis. Valve stenosis causes the valve flaps to thicken or stiffen, and they may fuse together. As a result, the valve opening narrows and blood flow through the valve decreases.

Atresia. The valve does not form in this condition, and a solid sheet of tissue blocks blood flow between the heart chambers.

3.2.1 Valvular heart disease- MRI

The advancements in CMR technology have resulted in a greater role for this modality in qualifying and quantifying various native valve diseases. Advances in CMR technology also enable higher spatial and temporal resolution imaging of various valves and their regurgitant or stenotic jets. As a result, CMRI can be an effective tool in assessing valvular heart disease. [1]

The utility of CMRI for valvular regurgitation evaluation was recently recognised as part of the American Society of Echocardiography's (ASE) and the Society of Cardiovascular Magnetic Resonance's (SCMR) joint recommendations for noninvasive valvular regurgitation evaluation (SCMR). Previously, CMR was only recommended in valvular disease when echocardiographic evaluation was insufficient or inconclusive. Cardiovascular magnetic resonance (CMR) has emerged in the last two decades as a noninvasive modality that does not use ionising radiation and is beneficial to patients with valvular heart disease. CMR imaging of valve anatomy allows for quantitative evaluation of stenosis and regurgitation. CMR can also detect the effects of the valvular lesion, such as ventricular volume or pressure overload and changes in systolic function. [2]

3.2.2 Valvular heart disease- ECG

ECG-gating is a technique for combining data from multiple consecutive heartbeats. As the most common finding on electrocardiography (ECG) in patients, LV hypertrophy is frequently associated with secondary repolarization abnormalities. This is found in 85 percent of severe patients. Its absence, however, does not rule out arterial stenosis (AS). Left atrial enlargement is common, as are conduction abnormalities such as left and right bundle branch block. This could be because the calcification has spread into the surrounding conduction system. The axis can be shifted left or right. Atrial fibrillation can also occur, particularly in the elderly and those with high blood pressure. [2]

3.2.3 Valvular heart disease- Echocardiography

Echocardiography is now the standard tool for the initial and long-term assessment of patients with valvular heart disease; however, echocardiography is

limited in patients with poor acoustic windows and may be more operator dependent than other modalities, particularly for disease severity quantification. [2]

Table -1: Classification of the severity of AS based on findings on Doppler echocardiography

	Aortic sclerosis	Mild	Moderate	Severe
Aortic jet velocity(m/s)	≤2.5	2.6-2.9	30-4.0	>4.0
Mean gradient (mm Hg)	-	<20 30a	20-40b 30-50a	>40b >50a
Aortic valve area (cm ²)	-	>1.5	1.0-1.5	<1.0
Indexed aortic valve area (cm ² /mm ²)	-	>0.85	0.60-0.80	<0.6
Velocity ratio	-	>0.50	0.25-0.50	<0.25

Doppler echocardiography is an excellent tool for assessing the severity of Articular Stenosis by measuring jet velocity and gradients as well as calculating aortic valve area. It also aids in the detection of other associated valve lesions as well as the estimation of pulmonary artery systolic pressure. [9]

3.2.4 Valvular heart disease- CT scan

Both electron beam and multislice cardiac computed tomography (CT) have been shown to correlate with echocardiographic assessment and clinical outcome. Although the role of CT in clinical management is not well defined, it is well established in evaluating the presence. [10]

3.3 Ventricular arrhythmia

Ventricular tachycardia (VT) is a leading cause of cardiac arrest. Patients with structural heart disease are more likely to develop malignant VTs. Multimodal imaging techniques are critical in determining the underlying cause of VT as well as its prognostic significance. Advances in imaging technology have allowed for more precise characterization of the structural arrhythmogenic substrate in patients with VT in recent years. Parallel to these advancements, the role of cardiac imaging has shifted from a primarily diagnostic tool to an

adjunctive tool for guiding interventional approaches for the treatment of VT.

3.3.1 VT- MRI

CMRI is superior in terms of accuracy and reproducibility in quantifying left ventricular ejection fraction (LVEF) and myocardial mass, and it can overcome the limitations of inadequate echocardiographic windows. CMRI is a one-stop shop for determining cardiac structure, function, and characterization of myocardial tissue. Late gadolinium enhanced (LGE) gadolinium CMRI imaging has become the gold standard for detecting myocardial fibrosis. In this method, gadolinium is used as a contrast agent to highlight areas of heterogeneity within the myocardium. Gadolinium washout is rapid in normal tissue but slow in areas of myocardial fibrosis. Normal and fibrotic tissue regions can be distinguished by timing image acquisition to occur "late," when washout has occurred in normal tissue but has not occurred in fibrotic tissue. This method works by changing the inversion time for distant normal myocardium to 'null,' causing it to appear black. Enhancement areas have been shown to correlate well with acute myocardial necrosis and chronic fibrosis in ischemic pathological specimens, as well as replacement fibrosis in non-ischemic dilated cardiomyopathy. [11]

3.3.2 VT- ECG

Depending on where the arrhythmia originates, ventricular tachycardia can have a variety of QRS morphologies, making ECG diagnosis difficult at times. Below are two examples of ventricular tachycardia with different QRS morphologies: one with a right bundle branch block morphology and one with a left bundle branch block morphology. In addition, VT can occur with any of the QRS morphologies in between. [12]

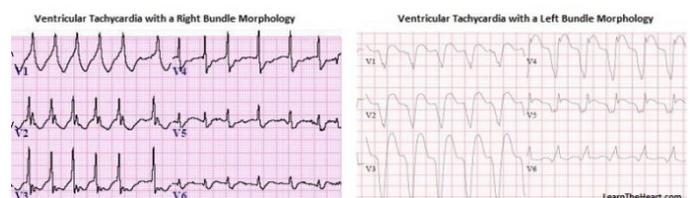


Fig -5: Ventricular Tachycardia with different QRS morphologies

3.3.3 VT- Echocardiography

The role of exercise echocardiography in asymptomatic AS patients is less clear due to a lack of research. Stress echocardiography also evaluates the LV functional response to exercise, with evidence indicating that patients with limited LV functional reserve (a lower increase in ejection fraction) are more likely to have an adverse event. More research is needed to determine whether an increase in the mean aortic valve pressure gradient and/or a limited increase in ejection fraction should be considered early elective surgery criteria. [9]

3.3.4 VT- MDCT (Multidetector CT)

MDCT has a limited role as a diagnostic tool in patients who present with VT. In circumstances where CMR and Transthoracic echocardiogram (TTE) imaging is unavailable or suboptimal, MDCT may also be considered as an alternative modality for detailed assessment of myocardial structure and function. Scar imaging using MDCT, as well as structural imaging, are currently primarily used as an adjunct during VT ablation procedures. [11]

3.4 ALCAPA Syndrome

ALCAPA syndrome, also known as Bland-White-Garland syndrome, is a rare congenital disease affecting one out of every 300,000 births and accounting for 0.25 to 0.5 percent of all congenital cardiac disorders. When an ALCAPA syndrome patient has a left-to-right shunt that causes abnormal left ventricular perfusion, the "coronary steal" phenomenon occurs. ALCAPA syndrome is one of the most common causes of myocardial ischemia and infarction in children. If ALCAPA syndrome is not treated, up to 90% of patients will die during their first years of life. ALCAPA syndrome can lead to myocardial infarction, left ventricular dysfunction, and mitral regurgitation in adults, as well as silent myocardial ischemia, which can lead to sudden cardiac death. Early detection and surgical intervention aimed at restoring a two-coronary-artery circulation system have great outcomes and lead to progressive myocardial healing. [13]

3.4.1 ALCAPA- ECG

Even though the ECG is generally abnormal, a normal ECG does not rule out ALCAPA. ALCAPA patients frequently have atypical Q waves in the lateral leads,

which are uncommon in normal newborns. In ALCAPA, Q waves in the inferior leads are infrequent. It's possible that a small Q wave disruption will be visible. When compared to adult ischemic heart disease, the Q wave is likewise unusual in that it is significantly deeper but narrower. [14]

3.4.2 ALCAPA- Echocardiography

The diagnosis of ALCAPA on echocardiography is usually straightforward, however even the most experienced echocardiographers can miss it occasionally. The majority of the time, children are referred with an echo lab to determine the cause of congestive heart failure. All young infants with ventricular dysfunction and regional wall motion abnormalities similar to those seen in myocardial infarction should be suspected of ALCAPA. The anterolateral LV myocardium is typically damaged and thinned down, while the posterior basal section may display hypertrophy and hyperfunction. Hyper echogenicity (scarring) of the papillary muscles, endocardial fibroelastosis of the left ventricle, and mitral regurgitation are visibly present. [14]

3.4.3 ALCAPA- MRI & CT scan

ALCAPA imaging findings in adults on CT and MRI include direct visualisation of the LMCA's origin from the posterior aspect of the pulmonary artery (PA), dilatation of the right coronary artery (RCA), and visualisation of dilated intercoronary collateral arteries along the external surface of the heart or within the interventricular septum. A subsequent contrast-enhanced CT of the chest revealed an enlarged LMCA that originated from the posterior aspect of the main PA, as well as a dilated and tortuous LAD and an enlarged tortuous RCA that originated orthotopically from the right coronary sinus of the aortic root. The coronary artery system as a whole seemed to be dilated and convoluted. Anatomic results on a confirmatory cardiac MRI were comparable. [15]

3.5 Wolff Parkinson White Syndrome:

Wolff-Parkinson-White syndrome (WPW) is a type of ventricular pre-excitation involving an accessory conduction route that affects 0.1 to 3% of the general population. Wolff Parkinson White Syndrome is thought to be caused by the presence of abnormal electrically conductive pathways between the atria and ventricles at birth (WPW). The disease includes auxiliary electrical channels that bypass the AV node.

3.5.1 WPW- Echocardiography

M-mode echocardiography can detect the tiny premature wall motion anomalies associated with WPW syndrome. However, pinpointing the exact location of the auxiliary pathway with sufficient precision is impossible. Even in patients with poor acoustic windows, different modalities of doppler echocardiography improve left-sided accessory route localization accuracy to 80-90 percent. [16]

3.5.2 WPW- ECG

A short PR interval (120 ms), a prolonged QRS complex (>120 ms), and a QRS morphology consisting of a slurred delta wave will be seen on the ECG. During typical sinus beats, a route could only transfer electrical impulses from the ventricle to the atrium and would not produce ventricular pre-excitation. Only an electrical impulse generated in the ventricle, such as a premature ventricular contraction or ventricular pacing, can reveal this hidden auxiliary circuit on ECG.

Young, healthy patients with no concomitant diseases or substantial risk factors who exhibit the WPW pattern on their ECG but are asymptomatic and have no history of suspected tachyarrhythmia are likely safe to follow up with primary care and/or cardiology. Since the WPW pattern alters the ECG's baseline morphology, the diagnosis of diseases based on ECG criteria may be affected in patients with the WPW pattern. [17]

3.5.3 WPW- CT scan

Through a consensus reading, the observers identified patients with cardiac anomalies. Observers reported aberrant observations in the apical, mid, and basal left ventricles for patients with WPW syndrome, using standard nomenclature for accessory pathway (AP). The wall thickness of the aberrant myocardium was measured at its thinnest point and compared to the remote normal myocardium from a short-axis, or four-chamber perspective. [18]

4. Conclusion

The survival of patients with congenital heart disease, whether treated or untreated, is expected to result in a large number of adults with congenital disease, and many more adult cardiologists will need to be trained to manage moderate to complex congenital lesions. Controversies in structured

abnormalities are detected by echocardiography in patients with brugada syndrome.

- Imaging technology advancements have enabled us to precisely detect structural arrhythmogenic substrates in VT patients.
- While echocardiography remains the primary method for visualising valve anatomy, CMR is a viable alternative if ultrasound windows are inadequate.
- Although diagnostics are run using ECG and Echocardiography, their normal reports do not rule out the presence of ALCAPA.
- The WPW pattern alters the baseline morphology of the ECG, which may affect the diagnosis of conditions that rely on ECG criteria in patients with the WPW pattern. [17]

5. Future Work

- Wolff-Parkinson White syndrome can be rarely detected by MRI.
- The role of CT in valvular heart disease has been well established yet the research done is negligible.

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REFERENCES

- [1] "Brugada Syndrome - NORD (National Organization for Rare Disorders)." <https://rarediseases.org/rare-diseases/brugada-syndrome/> (accessed Sep. 30, 2021).
- [2] K. Maganti, V. H. Rigolin, M. E. Sarano, and R. O. Bonow, "Valvular heart disease: Diagnosis and management," *Mayo Clin. Proc.*, vol. 85, no. 5, pp. 483-500, 2010, doi: 10.4065/mcp.2009.0706.
- [3] J. Derganc and G. Gomiscek, "Teaching the basic principles of electrocardiography experimentally," *Adv. Physiol. Educ.*, vol. 45, no. 1, pp. 5-9, 2021, doi: 10.1152/ADVAN.00155.2020.
- [4] O. Catalano et al., "Magnetic resonance investigations in Brugada syndrome reveal unexpectedly high rate of structural abnormalities," *Eur. Heart J.*, vol. 30, no. 18, pp. 2241-2248, 2009, doi:

- 10.1093/eurheartj/ehp252.
- [5] M. Takagi et al., "Localized right ventricular morphological abnormalities detected by electron-beam computed tomography represent arrhythmogenic substrates in patients with the Brugada syndrome," *Eur. Heart J.*, vol. 22, no. 12, pp. 1032–1041, 2001, doi: 10.1053/ehj.2000.2424.
- [6] A. Naseef, E. R. Behr, and V. N. Batchvarov, "Electrocardiographic methods for diagnosis and risk stratification in the Brugada syndrome," *J. Saudi Hear. Assoc.*, vol. 27, no. 2, pp. 96–108, 2015, doi: 10.1016/j.jsha.2014.06.004.
- [7] P. J. Cawley, J. H. Maki, and C. M. Otto, "Cardiovascular magnetic resonance imaging for valvular heart disease. Technique and validation," *Circulation*, vol. 119, no. 3, pp. 468–478, 2009, doi: 10.1161/CIRCULATIONAHA.107.742486.
- [8] R. C. Mathew, A. I. Löffler, and M. Salerno, "Role of Cardiac Magnetic Resonance Imaging in Valvular Heart Disease: Diagnosis, Assessment, and Management," *Curr. Cardiol. Rep.*, vol. 20, no. 11, 2018, doi: 10.1007/s11886-018-1057-9.
- [9] E. Picano, P. Pibarot, P. Lancellotti, J. L. Monin, and R. O. Bonow, "The Emerging Role of Exercise Testing and Stress Echocardiography in Valvular Heart Disease," *J. Am. Coll. Cardiol.*, vol. 54, no. 24, pp. 2251–2260, 2009, doi: 10.1016/j.jacc.2009.07.046.
- [10] C. J. Bennett, J. J. Maleszewski, and P. A. Araoz, "CT and MR imaging of the aortic valve: Radiologic-pathologic correlation," *Radiographics*, vol. 32, no. 5, pp. 1399–1420, 2012, doi: 10.1148/rg.325115727.
- [11] S. Mahida et al., "Cardiac Imaging in Patients with Ventricular Tachycardia," *Circulation*, vol. 136, no. 25, pp. 2491–2507, 2017, doi: 10.1161/CIRCULATIONAHA.117.029349.
- [12] Healio, "Ventricular Tachycardia (VT) ECG Review." 2020, [Online]. Available: <https://www.healio.com/cardiology/learn-the-heart/ecg-review/ecg-topic-reviews-and-criteria/ventricular-tachycardia-review>.
- [13] E. Peña, E. T. Nguyen, N. Merchant, and C. Dennie, "ALCAPA syndrome: Not just a pediatric disease," *Radiographics*, vol. 29, no. 2, pp. 553–565, 2009, doi: 10.1148/rg.292085059.
- [14] M. Varghese and S. Kothari, "The caveats in the diagnosis of anomalous origin of left coronary artery from pulmonary artery (ALCAPA)," *Images Paediatr. Cardiol.*, vol. 12, no. 3, p. 3, Jul. 2010, Accessed: Sep. 30, 2021. [Online]. Available: [/pmc/articles/PMC3228333/](https://pubmed.ncbi.nlm.nih.gov/2283333/).
- [15] J. Kothari, K. Lakhia, P. Solanki, D. Parmar, H. Boraniya, and S. Patel, "Anomalous origin of the left coronary artery from the pulmonary artery in adulthood: Challenges and outcomes," *Korean J. Thorac. Cardiovasc. Surg.*, vol. 49, no. 5, pp. 383–386, 2016, doi: 10.5090/kjtcs.2016.49.5.383.
- [16] A. Wu et al., "Enhanced Reader.pdf," *Nature*, vol. 388, pp. 1–14, 2020.
- [17] L. Chhabra, A. Goyal, and M. D. Benham, "Wolff Parkinson White Syndrome," *Card. Electrophysiol. Clin. Case Rev.*, pp. 267–269, Aug. 2021, Accessed: Sep. 30, 2021. [Online]. Available: <https://www.ncbi.nlm.nih.gov/books/NBK554437/>.
- [18] H. J. Lee et al., "Detecting regional Myocardial abnormalities in patients with Wolff-Parkinson-White syndrome with the use of ECG-gated cardiac MDCT," *Am. J. Roentgenol.*, vol. 206, no. 4, pp. 719–725, 2016, doi: 10.2214/AJR.15.15141.

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