Introduction

Sudden cardiac death (SCD) is defined as an unexpected sudden death due to cardiac cause and occurring within one hour of symptoms and is a major public health problem. SCD accounts for 30% of sudden deaths between 14 and 25 years of age. There is an increased risk of SCD in athletic young adults. There are a number of underlying cardiac conditions that increase the risk of SCD even though acute myocardial infarction as a correlate of cardiac cause is most common. Different underlying causes can be diagnosed and differentially identified by contrast-enhanced CT and MRI non-invasively.

Cardiac CT and MRI Findings of Structural Heart Diseases causing Sudden Cardiac Death in Adults

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Ischemic Heart Disease

- CAD with MI or angioplasty
- Myocardial infarction

Nonischemic heart disease

- Hypertrophic, dilated, restrictive
- Valvular heart disease
- Congenital heart disease
- Myocarditis
- Pericarditis
- Acute myocardial rupture
- Acute aortic dissection

No structural Heart Disease

- Primary electrical disease
- Prolonged QT syndrome
- Preexcitation syndrome
- Complete heart block

Chest wall trauma

Myocardial bridging

- Present in about 50% of normal hearts
- Most commonly middle segment of LAD
- Clinically silent in most cases
- Rarely, myocardial ischemia and infarction

Coronary artery disease with angiography or MI

- 80% of patients who experience SCD at all ages have atheroembolic coronary artery disease
- At postmortem acute occlusive coronary thrombosis was found in 1-64% due to plaque fissuring, hemorrhage or thrombosis
- No difference in artery involved
- More risk of SCD with abrupt proximal obstruction rather than progressive diffuse disease when collateral can form
- Patient prognosis with AMI depends on the extent of myocardial necrosis, presence of microvascular obstruction, and the degree of contractile dysfunction

Coronary artery dissection after blunt trauma

- Luminal, thrombosis, intimal dissection, arteriovenous fistula, pseudoaneurysm formation
- Coronary dissection consists of about 36% of traumatic coronary injuries
- MCDT: Ideal imaging technique to delineate intramural hematoma, assess luminal narrowing and follow up the appearance
- Useful to decide to do coronary angiography or not in patients with borderline probability of coronary injury

Anomalous coronary artery from the opposite sinus

- Risk of adverse events is highest in anomalous arteries with interarterial course
- RCA from left coronary sinus - 90%
- LCA from right coronary sinus - 60%
- LAD from right coronary sinus - 50%
- The cause of 5-35% of SCD in young athletes
- Mechanisms are not clearly
- Aberrant artery arises at an acute angle, often from a slit-like hypoplastic aorta, traverses the aortic wall obliquely, emerges between the aorta and the right ventricular outflow tract, and then proceeds to its usual area of distribution
- With exertion, proximal anastomosis or compression obstructs the blood flow, leading to dilated ischemic, ventricular tachycardia, and fibrillation

Coronary artery fistula

- An anomalous communication between a coronary artery and either a cardiac chamber, coronary sinus or pulmonary artery
- More commonly involving the RCA (60% of cases) than the LCA (40%)
- The commonest sites of drainage are the right ventricle (66%), right atrium (25%), or pulmonary artery (15%), producing left-to-right shunt
- If fistula flow is high, the distal perfusion of the originating coronary artery may be compromised

Myocardial bridging

- Coronary artery dives into the substance of the myocardium (intramyocardial course)
- Commonly middle segment of LAD
- Clinically silent in most cases
- Rarely, myocardial ischemia and infarction, arrhythmia, SCD
- Incidence: angiographic 0.5-3.5%, CT 3.5-30.5%
- Atherosclerotic plaque proximal to tunneled segment
- Angiography: milking effect and step-down step-up phenomena induced by systolic compression of MB segment
- MDCT: superficial and deep type, systolic compression

- More risk of SCD with abrupt proximal obstruction rather than progressive diffuse disease when collateral can form

Acute aortic dissection

- The most common of cardiomyopathy with an incidence of 1 in 500
- The commonest cause of SCD in the US (70% of patient suddenly die)
- Most common inherited cardiac disease (autosomal dominant)
- Basal anterior and anterior septal are most commonly affected
- Wall thickness > 30 mm infers an increased risk of SCD
- Systolic anterior motion (SAM) of the mitral leaflet with associated mitral regurgitation is frequent finding
- With increasing wall thickness, there is increased percentage of fibrosis. As fibrosis increases, wall function decreases

Hypertrophic cardiomyopathy

- Systolic anterior motion (SAM) of the mitral leaflet with associated mitral regurgitation is frequent finding
- With increasing wall thickness, there is increased percentage of fibrosis. As fibrosis increases, wall function decreases

Drowning

- Pickwickian syndrome
- Noncardiac Disease

- Acute aortic dissection
- Myocardial bridging
- Myocardial infarction
- Arrhythmogenic RV dysplasia
- Hypertrophic cardiomyopathy
- Drowning
- Pickwickian syndrome
- Noncardiac Disease
Non-ischemic dilated cardiomyopathy (NIDCM)

- Distortion of all four chambers of the heart, but primarily the left ventricle, with associated systolic dysfunction.
- 18% of symptomatic patients with ejection fraction less than 30% were diagnosed with NIDCM.
- Patients with NIDCM suffer from heart failure mortality and SCD in near equal numbers.
- SCD may well be the first manifestation of NIDCM, and idiopathic NIDCM is responsible for 10% of all SCDs in adults.
- It is often preceded by infectious myocarditis but 20-30% is familial. Other causes include autoimmune, toxic and metabolic diseases.
- A mid-myocardial stripe on delayed contrast-enhanced cardiac imaging is the “classic” imaging pattern seen in about 30% of patients. 60% of patients will have no enhancement.

Amyloid cardiomyopathy

- Almost all cases of primary amyloidosis and approximately 1% of familial amyloidosis.
- Amyloid deposition occurs with the myocardium of the atria and atrioventricular nodal atrophy and within the coronary arteries.
- Symptoms occur from disruption in contractile function, conduction disturbance and incompetence coronary artery flow.
- Left ventricular wall thickness - prognostic importance (measuring wall motion). Cardiac MR imaging: diffuse hypoenhancement pattern on delayed-enhancement MRE, typically more prominent in the subendocardium and the basal segments.

Non-compaction cardiomyopathy

- Rare cardiomyopathy which is believed to be a result of an arrest in development of the myocardium.
- Gross anatomic appearance: numerous, excessively prominent trabeculations and deep intertrabecular recesses in communication with the ventricular cavity.
- Predominantly (80%) found in the epicardial and mid-ventricular areas of both the inferior and the lateral wall.
- Eccentric hypertrophy, LV systolic dysfunction and heart failure (most common), arrhythmia, thrombembolism, SCD.
- Non-compacted to compacted ratio > 2.3 is diastolic dysfunctio assessed by MR criteria.
- Diastolic differential: prominent normal trabeculations, ARVD, apical HCM, thrombus, endocardial fibrosis

Arrhythmogenic right ventricular dysplasia

- Myocardial cell loss with partial or total replacement of right ventricular muscle and fat deposits occur in the late ventricular cavity.
- A familial disorder in 30% of cases, with autosomal dominant inheritance.
- The most common cause of SCD in an Indian screening program.
- Unexplained syncope on SCD is the first clinical manifestation of the ARVD.
- Onset in adolescence or early childhood.
- CT: Abnormal adipose connective tissue, prominent trabeculations with left atrophy, scalloped appearance of RV free wall, intramyocardial fat deposits.
- MRI: Intramyocardial fat, RV wall thinning or hypertrophy, trabecular dysrrhythmia of RV, enlargement of RVOT, global and regional RV dysfunction, myocardial fibrosis with delayed contrast enhancement

Myocarditis

- Most cases are subclinical.
- Most common cause: Group B streptococcus virus.
- SCD is a recognized complication and is often associated with exercise due to poor contractions of a dilated LV or arrhythmogenic focus from acutely inflamed or healed myocardium.
- MRI: Multiple patchy enhancement predominantly in subepicardial, interstitial portion.
- Follow-up: decreased extent of enhancement.

Aortic stenosis (AS)

- Most common valve disease resulting in valve replacement.
- Causes: Congenital (bicuspid aortic valve), degenerative, rheumatic heart disease.
- Pathophysiologic effects of AS
  - LV hypertrophy, LV systolic dysfunction, myocardial fibrosis, abnormal coronary blood flow, pulmonary hypertension.
  - Incidence of SCD in asymptomatic severe AS: 0.4-1%
  - Mechanism of SCD: VAD, and both malignant ventricular arrhythmias and bradycardia have been documented.
- Timing of surgery for AS
  - Symptomatic moderate to severe AS
  - Asymptomatic severe AS
  - Progressive increase in AV velocity and decrease in ARA
  - Critical AS (mean grad > 65 mmm, AVA 0.7 cm²).
- LV systolic dysfunction
  - Marked LHV
  - Associated severe coronary artery disease.

Mitral valve prolapse (MVP)

- “Billowing” or prolapse of one or both of the mitral leaflets into the left atrium.
- With a prevalent condition more associated with non- lethal arrhythmias.
- Whether simple MVP causes SCD is unclear as it is such a prevalent condition which may be coincidental finding and not necessarily SCD.
- Patients who also have mitral regurgitation and LV dysfunction or myocardial degeneration of the valve are at higher risk of SCD.
- ABC: cause of severe non- ischemic MR in the USA.
- Overall prognosis: excellent, but serious complications include endocarditis, SCD, severe MR.
- Pathophysiologic basis
  - Leaflet thickening and redundancy (myxomatous degeneration).
  - Electrocardiography
    - single or bileaflet prolapse at least 2 mm
  - Association with heritable connective tissue disorders
  - Marfan syndrome, Ehlers-Danlos syndrome, osteogenesis imperfecta, and pseudoachondroplasia.

Marfan’s syndrome

- Autosomally inherited connective tissue disorder occurring in about 1 in 10000 people.
- Defects in fibrillin-1 gene (FBN1) on chromosome 15.
- Clinical features
  - Longitudinal growth and weakness of the aorta which weakens it.
  - “Billowing” or prolapse of one or both of the mitral leaflets into the left atrium.
  - Non-compacted to compacted ratio > 2.3 in end-diastole.
  - Normally considered diagnostic by MRI criteria.
- Onset in adolescence or early childhood.
- CT: Abnormal adipose connective tissue, prominent trabeculations with left atrophy, scalloped appearance of RV free wall, intramyocardial fat deposits.
- MRI: Intramyocardial fat, RV wall thinning or hypertrophy, trabecular dysrrhythmia of RV, enlargement of RVOT, global and regional RV dysfunction, myocardial fibrosis with delayed contrast enhancement.

Aortic dissection

- Longitudinal cleavage of the aortic media by dissecting column of blood.
- Almost always accompanied by a transverse innal and medial tear.
- Intimal tear sites: Type 1 and II – > 2 cm above the sino-tubular junction involving right lateral wall.
- Type IIIB (distal to the origin of the left subclavian artery).
- False lumen in classic dissection.
- Within the media:
  - Intimal flap: inner 2/3 of media + intima.
  - Complication: rupture, leakage, side branch involvement, aortic occlusion, dissection aneurysm.

Conclusion

1. SCD is not common in the general population. However, the incidence is significantly higher in the cohort of patients who has general cardiac disease. The causes of SCD in adults vary with age.
2. It is very important for the radiologists to be familiar with radiological findings of the predisposing underlying cardiac abnormalities may allow preventive management.