Cardiovascular System

Symptoms

Sudden death

Preventing sudden death is the highest priority in care of people with DM1. Sudden cardiac deaths in DM1 are mostly attributable to complete cardiac conduction block and ventricular fibrillation/tachycardia caused by cardiomyopathy.

Syncope and presyncope

Cardiogenic syncope should be first considered in management of patients with DM1. Cardiogenic syncope and presyncope may precede a sudden cardiac death. Milder complaints, such as non-vertiginous dizziness and lightheadedness, should also be considered as potential cardiogenic events.

Cardiac conduction defects

While patients with severe cardiac conduction block may present with the above-mentioned symptoms, patients with milder conduction blocks may be asymptomatic, especially when the conduction block does not cause significant hemodynamic changes. However, conduction delays at the AV node, the His bundle, and within the ventricle should be carefully assessed for indications of potential interventions.

Cardiac arrhythmias

The most common type of arrhythmia in patients with DM1 is atrial fibrillation/flutter, which poses risks for cardiogenic embolism. Various tachyarrhythmias and bradyarrhythmias are often symptomatic and may cause palpitations, fatigue, chest pressure or pain, dyspnea, syncope, presyncope, lightheadedness and dizziness. A high-degree AV block should be first considered as a possible cause of bradycardia in DM1 patients. Episodes of ventricular and supraventricular tachyarrhythmias may cause syncope or presyncope.

Hypotension

Hypotension is often found in patients with DM1 or DM2. Although hypotension has been attributed to autonomic dysfunction, the exact mechanism remains unknown.

Congestive heart failure

Dilated cardiomyopathy may lead to congestive heart failure in advanced stages of the disease. Pulmonary hypertension often leads to cor pulmonale in neonates born with congenital myotonic dystrophy DM1.

Adult myotonic dystrophy DM1 patients usually (but not always) develop cardiac manifestations after developing neuromuscular symptoms. Some asymptomatic children with DM may be at risk for sudden cardiac death.

Patterns Of Cardiovascular System Problems

Congenital DM1
Newborn • Pulmonary hypertension and cor pulmonale • Cardiomyopathy in rare cases
Childhood/Adolescence • Possible early cardiomyopathy and cardiac conduction problems
Adulthood • Dilated cardiomyopathy and cardiac conduction defects beginning in early adulthood. These heart problems are one of the main causes of early mortality seen in adult patients with congenital myotonic dystrophy DM1.
Childhood Onset DM1
Childhood/Adolescence • Possible cardiomyopathy and cardiac conduction problems beginning in the second decade
Adulthood • Dilated cardiomyopathy and cardiac conduction defects sometimes present beginning in early adulthood • Complications in severe cases can lead to heart failure and sudden death, even in asymptomatic individuals. • Hypotension
Adult Onset DM1
Hypotension • Dilated cardiomyopathy and cardiac conduction defects possible. Complications in severe cases can lead to heart failure and sudden death, even in asymptomatic individuals.
DM2
Hypotension • Mild arrhythmia and other conduction issues occasionally present • Dilated cardiomyopathy and cardiac conduction defects possible, but less common than DM1. Complications in severe cases can lead to heart failure and sudden death, even in asymptomatic individuals.

Diagnosis

- Annual cardiological history and physical examination
- Annual 12-lead electrocardiogram (EKG)
- 24h portable Holter monitor if symptoms suggest cardiac arrhythmias or cardiogenic syncope, or if EKG shows cardiac arrhythmias or conduction abnormalities
- 2D / M-Mode Echocardiography every 2-5 years

- Invasive electrophysiology (EP) testing when potential for serious conduction blocks or arrhythmias are suspected. Because of the possibility of sudden death, the EP testing should be performed with relatively liberal indications.

Treatment

Cardiac Devices

The use of implantable cardiac pacemakers and cardioverter defibrillator devices may be warranted, depending on EP results. Due to the potential for sudden, rapid symptomatic progression and recurrent cardiac events, patients with DM are considered to have a class I indication for cardiac pacing with second and third degree AV block, and a class IIb indication for cardiac pacing even with first degree AV block, regardless of symptoms. (See Europace 2007 9(10):959-998 for guidelines on cardiac pacing and cardiac resynchronization therapy). However, some debate exists regarding the use of these devices, as their utility has not been established for all patients.

Medications

Anti-arrhythmic drugs are available for individuals with milder symptoms. However, Class I anti-arrhythmic drugs are contraindicated as they may have pro-arrhythmic effects. Sudden vigorous exertions should be avoided since sudden death has been associated with rapidly elevated heartbeat. Congestive heart failure should be managed with conventional treatments.

The cautious use of anti-myotonic medications and general anesthetics is also warranted, as they can elevate the risk of cardiorespiratory complications.