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Anti-arrhythmic and anti-inflammatory effect of low-level electrical stimulation of aortic root ventricular ganglionated plexi in dogs with heart failure

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Background & Aim: Heart Failure (HF) and arrhythmia often coexist and share the similar underlying pathogenesis, including autonomic imbalance, electrical remodeling and inflammatory reactions. Low Level-Electrical Stimulation (LL-ES) rebalances the tone of the autonomic nervous system and has an anti-arrhythmic effect. However, it is unknown whether LL-ES can decrease the inflammatory response and benefit patients suffering from both HF and arrhythmia. This study aimed to investigate the anti-arrhythmic and anti-inflammatory effects of LL-ES of Aortic Root Ventricular Ganglionated Plexi (ARVGP).

Method: 20 dogs were divided randomly into drug administration (control) and LL-ES groups after performing rapid right ventricle pacing to establish the HF model. The inducing rate of arrhythmia was measured after a programmed electrical procedure at the baseline and drug administration or LL-ES. The bioactive factors of HF, including angiotensin II, TGF-β, Mitogen-Activated Protein Kinase (MAPK) and Matrix Metallo Proteinase (MMP), were assessed. Furthermore, ventricular size and left ventricular ejection fraction were determined.

Result: Compared with the control group, the inducing rate of arrhythmia decreased from 40% to 10% after 4 h of LL-ES ($P<0.05$). The expression of angiotensin II, TGF-β, MAPK, and MMP was down regulated significantly in the LL-ES group ($P<0.05$). Moreover, the volume of the left ventricle and the ejection fraction of the left ventricle in the LL-ES group changed little ($P>0.05$).

Conclusion: Short-term LL-ES of ARVGP presented both anti-arrhythmic and anti-inflammatory effects and contributed to the treatment of HF and the associated arrhythmia.

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Cardiac rehabilitation: The emerging role of home-based approaches and telemedicine

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Outpatient cardiac rehabilitation programs provide supervised exercise training in addition to secondary prevention interventions. They are designed to speed recovery from acute cardiovascular events, benefit chronic patients and to improve quality of life. Alternative approaches to the delivery of supervised cardiac rehabilitation include home-based programs, disease management and lifestyle health coaching interventions and other internet-based case management systems. The effectiveness of home-based programs was evaluated in several randomized trials. There was no evidence of a difference in mortality, re-infarction, revascularization, cardiac-associated hospitalization or exercise capacity between the two modes of intervention. Other alternatives include community-based group programs and the use of telemedicine. Use of mobile health technologies may further expand cardiac rehabilitation availability. Telehealth exercise cardiac rehabilitation appears to be at least as effective as center-based cardiac rehabilitation in improving modifiable cardiovascular risk factors and functional capacity. It identifies the option of telehealth and the technologic advances to provide more comprehensive, responsive and interactive interventions for individuals for whom center-based rehabilitation is not feasible. The attractiveness of telemedicine models is the potential to improve participation of patients in structured with its short term and long-term benefits.

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Rare congenital disorder and heart failure complex surgical repair and post-operative regeneration in an adult female

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The World Health Organization Global Hearts initiative acknowledges Cardiovascular Disorders as the number one killer in the world. It also recognizes the paucity of information and understanding about cardiovascular disorders, particularly as regards women with adult Congenital Heart Defects (CHD), on right heart failure, Transposition of the Greater Vessels (TGV) the high incidence in the developing world, the potential impact of travel and exposure to prolonged seated immobility, and the journey through diagnosis, treatment and post-surgery recovery challenges.

Through presentation of this incredible journey to symptom manifestation, diagnosis, surgical repair and the equally incredible post-surgery recovery period of heart and body regeneration from this particularly rare compound of congenital defects that surfaced only near mid-life in an adult female, this case story may explode some commonly-held notions of CHDs and illuminate directions for further diagnosis and treatment in general and in relation to adult women in particular. This case of right heart anomalies and failure also point to some startling departures in popular understanding of heart conditions largely drawn from knowledge of left heart anomalies.

Additionally, in a rare zone of survival, it highlights personal efforts to treat with the seemingly little understood post-operative period of a body and its organs readjusting to restructured heart after a lifetime, coping with regeneration and its survival efforts to date. It further aims to highlight some of the challenges and limitations of screening and detecting, particularly in the context of health care in the developing world.

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The role of ultra-high density mapping system to unveil hidden signals: A case report

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Background: Atypical Atrial Flutter (AAFL) is often associated with structural heart disease, especially in patients that have undergone cardiac surgery or extensive catheter ablation for the treatment of AF (Atrial Fibrillation).

Method: We present the case of a 60-year-old female with history of multiple AF ablations referred to our hospital for symptomatic persistent AAFL that was finally eliminated using a novel Ultra-High Density (UHD) mapping system. The 3D atrial geometry was created from the basket mapping catheter IntellaMap ORION™ and the RHYTHMIA™ mapping system. A trans-septal access was carried out in order to complete the electro-anatomic map of the left atrium.

Result: The activation map revealed an isthmus of slow conduction localized along the inferior part of the left inferior pulmonary vein. Analysis of EGM potentials revealed a concordance with the meso-diastolic activation. Interesting, the appearance of these potentials were detectable only on Orion basket whereas the ablation catheter did not revealed any sign of atrial activation. A single shot radiofrequency application of less than 60 sec along the critical isthmus channel promptly interrupted the arrhythmia and the sinus rhythm was restored. Patient remained free from AAFL recurrence and symptom after 6-month follow-up.

Conclusion: In this case the small, close, and low-noise mini-electrodes of orion catheter allow us to identify the critical isthmus of arrhythmia (represented by an extremely low voltage meso-diastolic potential) completely not recognizable by the ablation catheter. UHD mapping seems to be of a primary importance in patients with substrate-related atrial tachycardia and previously failed ablation.

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FMTVDM©℗: A quantum leap forward for the fields of nuclear cardiology and nuclear medicine

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Background: The foundational work of nuclear cardiology and nuclear medicine began with Blumgarts 1925 study of circulation time. The method was actually quantitative yielding measurements of isotope over time. Unfortunately, the field of nuclear medicine and later nuclear cardiology would yield to an approach of qualitative image interpretation resulting in problems with sensitivity and specificity as do all qualitative methods, resulting in a 35% error rate, matching the limitations of anatomic assessment of disease, including but not limited to coronary angiography, mammography, CT/MRI, etc.

Method: 300 men and women between ages 21 and 85 years of age were studied in five centers across the US, using a quantitative and enhanced method (FMTVDM©℗) designed to measure isotope (Sestamibi and Myoview) redistribution to define wash-in, washout and normal redistribution.

Result: Results were compared to Quantitative Coronary Angiography (QCA). Using FMTVD redistribution measurements, percent Diameter Stenosis (%DS) was then calculated and the calculated %DS used to calculate a quantified/Fleming coronary flow reserve© then used to calculate coronary artery narrowing (%DS) and QCFR/FCFR using the proprietary patent equations. The resulting strong relationship for the coefficient of determination was 0.87582 ($p<0.0001$).

Conclusion: Qualitative comparisons of nuclear imaging produces a diagnostic error rate of 35% comparable with angiographic errors in reader interpretation and the inability to satisfactorily unmask underlying Vulnerable Inflammatory Plaques (VIPs) responsible for roughly 85% of all myocardial infarctions. FMTVDM©℗ provides the first ever quantified and enhanced method for measuring Coronary Artery Disease (CAD) beginning with the measurement of isotope redistribution and ending with the calculation of QCFR/FCFR© using the patented proprietary equations. This patented method is applicable to any device capable of measuring isotope activity over time including but not limited to hand-held probes, planar, SPECT (Single-Photon Emission Computed Tomography) and PET (Positron Emission Tomography). This provides the first quantitative and evolutionary change for the fields of nuclear medicine and nuclear cardiology since its inception in 1925, (QCFR/FCFR) using proprietary equations. The result was then compared with the QCA derived measurements using best fit regression analysis.

Results: FMTVDM©℗ measurements of Sestamibi and Myoview redistribution produced a parabolic relationship ($p<0.01$) and showed that both technetium 99-m isotopes redistribute beginning at 5-minutes post isotope infusion compared with the 60-minute distribution of isotope. Failure to correctly identify this timing of isotope redistribution had resulted in prior erroneous assumptions that Sestamibi and Myoview did not redistribute.

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Different techniques of cardiac leads extraction: 17-years single center experience

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Background & Aim: Cardiovascular Implantable Electronic Devices (CIEDs) have become an important therapeutic modality of cardiovascular care. The increasing rate of CIEDs implantation determines an increasing rate of CIEDs related complications, especially including devices and leads infections that represent the strongest indication for the complete system removal. Therefore, it is clear the importance of providing safe and efficient Transvenous Lead Extraction (TLE) techniques. TLE techniques used until now, although very effective, are not without risk especially in elderly patients with significant comorbidities. The aim of our study is to evaluate the safety and the efficacy related to lead extraction of CIEDs with all currently available techniques (simple manual traction, mechanical sheaths and high-frequency excimer laser). We explore the indications, complications, and success rates involved in the removal of CIEDs leads in our center, over 17 years.

Method: We enrolled 447 consecutive patients (mean age 72,8 years, 74% male and 26% female) undergoing TLE between December 2001 and February 2018. In total 955 leads were extracted, 353 of which were atrial, 278 ventricular, 213 ICD (Implantable Cardioverter Defibrillator) leads and 111 were left leads for coronary sinus. The indications for TLE were pocket infection (44%), sepsis (31%), lead failure (16%) and others causes (9%). Lead implant mean duration was 54 months. 87 leads (9%) were extracted using Lead-Locking-Device (LLD), 497 (52%) using high-frequency excimer laser, 235 (25%) using mechanical sheaths and 136 (14%) using simple manual traction. For 15 leads (1.5%) TLE needed femoral transvenous approach. A complete transvenous lead extraction without complications was considered successful.

Result: Overall 879 leads (92%) were successfully and completed extracted, 65 leads were partially extracted with no other complications and 11 leads (1%) were not extracted. There were major complications in 14 patients (3.13%) such us pericardial effusion, cardiac tamponade, bradycardia, superior cava vein rupture, sub-clavian vein thrombosis, jugular thrombosis and hematoma. All-cause mortality related to TLE procedure was 0.89% (n=4) at 1 year of follow-up. The all-cause mortality at two years of follow-up was 9.43% (n=45).

Conclusion: According to our experience, transvenous lead extraction can be safely and successfully performed in the majority of patients regardless of the technique used (LLD, high-frequency excimer laser and mechanical sheaths). However, the mortality of patients extracted for systemic infection remains slightly high at two years follow-up.

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A heart or a boot?

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A 20-year-old refugee arrived at our emergency department for the presence of recurrent episodes of breathlessness, palpitation and easy fatigue since childhood. In his country of origin he had never undergone investigations. His family history was devoid of important and did not take any medication. The triad composed of exercise dyspnea, easy fatigue and palpitation suggested a cardiac cause, and the presence of symptoms since childhood indicated that it was a systemic disease, although it could also be a rheumatic endocarditis. It showed signs of chronic malnutrition (growth retardation) and acute (body mass hypotrophy). It was plethoric, cyanotic and with severe digital hippocratism, signs that could be secondary to a congenital cyanogenic heart disease such as the tetralogy of Fallot or Eisenmenger's syndrome. The cardiovascular objective examination revealed a pansystolic murmur at the left inferior sternal angle, associated a thrill: findings present in both tricuspid deficiency and interventricular septal defects. However, the absence of cannon-shot waves made tricuspid insufficiency unlikely. The second cardiac tone was intense, suggesting the presence of pulmonary hypertension. On the other hand it was single and this indicated a pulmonary stenosis. The chest radiograph revealed oligemic lung fields and a heart shaped like a boot. At the EKG (Electro Cardio Graphy) pulmonary P-waves, right axial deviation and right bundle branch block were evident. The echocardiogram, which revealed a taking 2 cm defect of the membranous portion of the interventricular septum with a right-left shunt; knight aorta with normal aortic diameter; right ventricular hypertrophy, right atrial dilatation and infundibular stenosis, small left ventricle; 56% ejection fraction. At this point, the combination of cyanosis, interventricular defect and pulmonary stenosis was characteristic of the tetralogy of Fallot. In cases like this, the surgical correction must be performed as soon as possible. The boy was indeed transferred to cardiac surgery, where he continued the therapeutic process, which was successful.

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Predicting factors of adverse outcomes in STEMI patients undergoing primary and rescue percutaneous coronary intervention: Thailand tertiary care center experience

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Aim: The aim of this study was to determine the predicting factors of adverse events among STEMI patients undergoing primary and rescue PCI in Thailand.

Method: For the Initial outcome, Some STEMI (ST-Elevation Myocardial Infarction) patients was reviewed from STEMI charts in Hatyai Hospital. As per the Review from 11th February 2014 to 31st March 2017, review from OPD chart and IPD chart, the primary outcome was the predictors of adverse events among STEMI patients undergoing primary PCI (Percutaneous Coronary Intervention) and rescue PCI. Secondary outcome includes mortality, MACE (Major Adverse Cardiac Events), cardiac death, target vessel related re-infarction and ischemic driven target-lesion revascularization, stroke, recurrent myocardial infarction, heart failure and so on.

Result: The strongest factor of adverse events among STEMI patients undergoing primary PCI and rescue PCI is Killip class, the older age.

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Heart failure: Management failures, who is to be blamed?

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Heart Failure (HF) remains a major public health problem that has high incidence and prevalence globally. It is the leading cause of hospitalization for people of 65 years of age and older and rates of hospital readmission within 6 months range from 25% to 50%. The personal burden of HF includes debilitating symptoms, frequent re-hospitalizations and high rates of mortality. HF also poses a substantial economic burden, with annual direct costs for the care of HF patients estimated to be between $20 billion and $56 billion. A number of studies have documented marked variation in the quality of care judged by specific performance measures and substantial underuse of evidence-based, guideline-recommended, HF therapies in patients receiving conventional care. Moreover, patient behavioral factors (such as non-adherence to diet and medications) and economic and social factors frequently contribute to re-hospitalizations. The traditional model of care delivery is thought to contribute to frequent hospitalizations because in these brief episodic encounters, little attention may be paid to the common modifiable factors that precipitate many hospitalizations. Patient education, discharge plan, follows up and management at community level are variable and sub-optimal. Limited or poor patient participation and involvement in self-care is also a major factor in leading to poor outcome in HF. As such, there has been much interest in identifying effective methods to improve the quality of care for HF patients while reducing costs. An effective management strategy and a balanced approach is the much needed.

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MicroRNA-22 promotes the osteogenic differentiation of valvular interstitial cells by targeting calcium binding protein 39

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Calcific Aortic Valve Disease (CAVD) is a complex pathological process for which no effective therapies currently exist. Transformation of Valvular Interstitial Cells (VICs) to osteoblasts is believed to be one of the most important causes of valve calcification. Recently, emerging evidence suggests that pro-osteogenic MicroRNAs play essential roles in the calcification of the aortic valve. The purpose of this study is to determine whether miR-22 is critically involved in the osteogenic differentiation of VICs and if so, to determine the molecular mechanisms involved. A total of 33 CAVD patients were enrolled in the study. The severity of CAVD was determined by standard echocardiographic methods. To identify the aberrant expression of miRNAs in calcified aortic valve, real-time PCR was performed to detect the expression profiles of osteogenic miRNAs in CAVD patients. Subsequently, we identified miR-22 as one of the most significantly up-regulated miRNAs in calcified aortic valves. Fluorescence *in situ* hybridization assay showed that miR-22 was expressed throughout the regions of the calcified valves and predominantly localized in VICs, as indicated by the co-expression of vimentin. Elevated miR-22 levels were positively correlated with the expression of OPN ($rs=0.820$, $P<0.01$) and Runx2 ($rs=0.563$, $P<0.01$) as well as VIC osteogenic differentiation. Furthermore, we identified calcium binding protein 39 (CAB39) as a novel downstream target of miR-22 in VICs, as determined by dual-luciferase reporter assay, real-time PCR (Polymerase Chain Reaction) and western blot assays. Furthermore, we found that the CAB39 expression was negatively correlated with the calcification severity in clinical CAVD samples, as determined by immunohistochemical staining analysis. Adenovirus-mediated both gain- and loss-of-function analyses demonstrated that miR-22 is critically involved in the osteogenic differentiation of VICs, specifically through regulating the CAB39-AMPK-mTOR signaling pathway. MicroRNA-22 serves as a potential inducer of CAVD through inhibiting the CAB39/AMPK/mTOR signaling pathway. These results suggest that miR-22 may serve as a potential therapeutic target for the calcific aortic valve disease.

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Coronary artery aneurysms: Variable presentations and management

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Background: Coronary Artery Aneurysms (CAA) are rare clinical entities. They are found in 0.3-5% of patients undergoing coronary angiography. Angiography is the mainstay for diagnosis. The optimal management of CAA remains controversial and largely based on case reports and anecdotal experience.

Case Description: We report two cases of CAA. First, a 74-year-old man with history of typical angina pectoris underwent invasive coronary angiography that showed in the proximal Left Anterior Descending (LAD) a large saccular aneurysm. The second patient is a 78-year-old man presenting to the emergency department for chest pain with ST segment elevation. Coronary angiogram was emergently performed which showed Pseudo Aneurysm (PSA) of the distal right posterior descending coronary artery. Cardiac CT confirmed PSA and contained coronary perforation with hematoma in the pericardial space, suggestive of leaking aneurysm.

Case Outcomes: Cardiothoracic surgery performed surgical excision of the saccular aneurysm in the first patient with a left internal mammary artery bypass to the LAD (Left Anterior Descending). Two days after the procedure, the patient developed ventricular tachycardia that became refractory to medical therapy and the patient expired. The second patient had PCI (Percutaneous Coronary Intervention) with successful coil embolization of the PSA with no complications and was discharged home.

Discussion: Untreated CAA may be complicated by myocardial ischemia or infarction, distal embolization due to thrombus formation within the aneurysm and spontaneous rupture. The prognosis and management are controversial. Our two cases illustrate the varied presentations, management and outcomes in this rare patient population. More data and outcomes studies are needed to help guide future management of these patients.

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Acute limb ischemia: A case report

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Introduction: Acute Limb Ischemia (ALI) is a sudden decrease in limb perfusion that may threaten limb viability. The most common cause is acute arterial total or near-total occlusion. Mortality and morbidity rate were high even after surgical or endovascular intervention. Early recognition and revascularization are essential. A case of ALI is presented to illustrate the diagnostic and therapeutic approach.

Case Report: A 54-year-old man had history of ascending colon adenocarcinoma status post right hemicolectomy, sick sinus syndrome and complete atrioventricular block status post permanent pacemaker implantation, hypertension, type 2 diabetes mellitus and coronary artery disease. He presented with acute onset of numbness, pain and weakness of the right leg. Vital sign in emergency department revealed blood pressure 217/88 mmHg, body temperature 36.5 ºCelsius degree and heart rate 91/min. Physical examination revealed weak pulsation of right dorsal pedis artery and swelling and erythema of the right leg. Electrocardiography showed atrial fibrillation and pacemaker ventricular pacing rhythm. Laboratory data revealed leukocytosis and hyperglycemia. Computed tomography of the right lower extremity demonstrated an acute thrombosis with total occlusion from right Common Femoral Artery (CFA) to Superficial Femoral Artery (SFA).The patient underwent endovascular therapy with balloon angioplasty and thrombus fragmentation. The flow of SFA was restored after angioplasty but distal embolization of thrombus was noted to popliteal artery. Consequently, a multi-hole infusion catheter was placed over popliteal artery. Intra-arterial infusion with Urokinase 1,000,000 units for 24 hours and Enoxaparin were administered. His right lower limb became warm during therapy and his symptoms improved. Angiography was repeated on the next day. Flow of popliteal artery was restored. Mild reperfusion tissue swelling of the right leg was observed. Finally, the patient was discharged with non-vitamin K anti-coagulant Rivaroxaban.

Conclusion: ALI is a critical condition with high mortality and morbidity rate. Early recognition and early revascularization are essential. Endovascular treatment is one minimally invasive procedure with effect and quick response.

Discussion: ALI is related to high possibility of limb amputation, severe complication such as reperfusion injury and mortality. It is essential to achieve early recognition and diagnosis. Revascularization in viable case as soon as possible plays the major role in limb rescue and complication prevention. Surgical intervention and endovascular treatment are effective choices with quick response. Available treatment must be chosen rapidly in this emergent disease. In our case, endovascular therapy was decided. Blood flow of the acute ischemic limb was restored effectively with the minimally invasive procedure.

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Value of global longitudinal peak systolic strain derived by 2D speckle tracking in detection of obstructive coronary artery disease

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Background & Aim: Non-invasive identification of patients with Coronary Artery Disease (CAD) remains a clinical challenge despite the widespread use of imaging and provocative tests and speckle tracking echocardiography has been validated for assessment of global and regional left ventricular myocardial function which is affected in patients with obstructive CAD. The study aims the early detection of obstructive coronary artery disease using peak systolic global longitudinal strain derived by 2D speckle tracking in patients with chronic stable angina.

Method: 75 patients with chronic stable angina were enrolled in this cross sectional study, (Mean age was 56.69±6.96 y, 35 were males), 42.7 % were diabetic and all patients were assessed by thorough history taking, clinical examination, 12 lead surface ECG, conventional, speckle echocardiography and coronary angiography in Mansoura specialized medical hospital over a period of 7 months from march 2017 to October 2017.

Result: Statistically significant decrease was found in GLPS-Avg (Global Longitudinal Systolic Peak Strain) values in patients with obstructive CAD when compared to patients with normal coronary angiography ($p<0001$) and in patients with 3 or more risk factors when compared to patients with one or two risk factors ($p=0.014$) and when syntax score was increasing among patients with obstructive CAD a significant decrease in median GLPS-Avg values was noted ($p<0.001$), but when regional systolic strain values were compared to affected coronary arteries no significant difference was found ($p=0.844$) i.e. almost identical correlation between affected segments by speckle tracking and obstructed arteries by coronary angiography. Multivariate logistic regression analysis showed that GLPS-Avg was found as a predictor for obstructive coronary artery disease in patients with chronic stable angina ($p=0.028$ with odds ratio 31.4 and 95% CI (1.85-535)). ROC curves were established and cutoff value was determined for GLPS-Avg as -16 with 89.8% sensitivity and 100% specificity.

Conclusion: longitudinal strain derived by speckle tracking can be used as non-invasive simple test for evaluation of patients with chronic stable angina and as a predictor for presence or absence of obstructive CAD.

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Mutations in exons 30 and 33 of the MYBPC3 gene and its effect on hypertrophic cardiomyopathy

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Cardiomyopathy is a different category of heart muscle diseases that the heart cannot supply adequate blood flow for general and specific circulation of other organs of the body in which case the patient gets an arrhythmia. This disease has several types including Hypertrophic Cardiomyopathy (HCM), Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC), isolated non-compaction, mitochondrial cardiomyopathies, Acquired agents, secondary metabolic factors, inflammation, endocrine, poisoning and neuromuscular. This article mainly intended to explore new therapies by examining one of the genetic causes of this complication. The process of translating and expressing proteins exon and intron play a major role. Exons are nucleotide sequences that they are coded in the form of RNA and they have an important usage in the translation process for make essential proteins. In many genes existing exons have translatable areas that they lead to protein synthesis.one of effective genes in the process of completing the muscular process of the heart, considering the genetic cases that was said, is the MYBPC3 gene that mutations in exons 30 and 33 of this gene lead to a series of major changes in the development of myocardial heart muscles that its result is hypertrophic cardiomyopathy. Mutations in 23 Sarcomere genes or sarcomere-related proteins are related to HCM. Mainly changes in MYH7 and MYBPC3 genes that they are encoder of myosin heart binding protein C are the causes of more than 75% of all HCM clinical cases. This mutation in thin strand proteins such as troponin T, troponin I and tropomyosin include less than 10% of HCM cases. These observations indicate that created mutations cause changes in structure and performance of a mutated protein and common genetic variants in it and ultimately a genetic cardiomyopathy disorder.

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Brugada syndrome and its relation to mitochondrial cardiomyopathies

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Cardiomyopathy is a progressive myocardial disease or heart muscle. In most cases, the heart muscle weakens and the ability to pump blood to other parts of the body decreases. Cardiomyopathy can lead to irregular heartbeat, heart failure, disorders and diseases of the heart valves or other complications. Some effective measures are there to resolve this problem. Cardio myopathy has many causes that the most common one is the heart muscle is stretched and due to this stretching, the blood pumping become very weak. In the meantime, some nucleotide changes in molecular level at the mitochondrial are led to Brugada syndrome that one of the genetic reasons for sudden cardiac deaths is as a result of ventricular fibrillation. At the molecular level mutations that occur in the SCN5A gene that it is encoding the alpha subunit of the sodium channels of the heart cells, they lead to this syndrome. Any disorder that occurs in the mitochondrial respiratory chain leads to a defect in the function of the target tissue. As you know, the heart is one of the organs that needs a lot of energy to function and energy deficiency in cells affects ion channels including this complication cause heart disorders. Cytochrome C is part of the mitochondrial transduction of electrons and respiratory chain that it reconstructs H_2O and the nucleotide defects that occur in this mutation, it encompasses a wide range of diseases including myopathic disturbances to severe multi-systemic diseases. In accordance with the provided explanations, there is a direct correlation between the disturbances of the sodium channel that they play an important role in action potential of cardiac cell and this syndrome and created mutations cause to decrease the function of sodium channels in the heart muscle and it prevents the precise function of the blood supply system, because the heart is heavily dependent on the oxidative energy produced by the mother's inherited mitochondria.

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