4-1-2022

A Case of Muscular Dystrophy with Dilated Cardiomyopathy: Do Not Forget Your Basics

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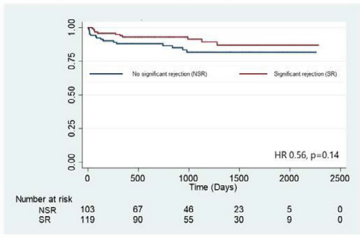
frequency routine surveillance EMB in post-HT patients, even in the early period when non-invasive diagnostics are not applicable.

Table 1
Hemodynamic measurements and LVEF from the first four post-transplant biopsies in 150 patients, 555 total biopsies

Biopsy Grade	N	RAP (mmHg)	RVSP (mmHg)	PASP (mmHg)	PADP (mmHg)	mPAP (mmHg)	PCWP (mmHg)	CO (L/min)	CI (L/min/m^2)	LVEF (%)
0/0R	319	7	33	32	13	21	13	5.5	2.9	69
1A/1R	120	7	33	30	13	20	12	5.3	2.7	68
2/1R	57	7	32	32	13	20	13	5.2	2.8	67
3A/2R	59	7	33	31	13	21	13	5.6	2.8	68

Data are presented as means. RAP: Right atrial pressure; RVSP: Right ventricular systolic pressure; PASP: Pulmonary artery systolic pressure; PADP: Pulmonary artery diastolic pressure; mPAP: Mean pulmonary artery pressure; PCWP: Pulmonary capillary wedge pressure; CO: Cardiac output, determined by thermodilution; CI: Cardiac index, determined by thermodilution; LVEF: left ventricular ejection fraction.

Figure 1
Kaplan-Meier post-transplant survival analysis for patients with no significant history of rejection versus those with significant rejection



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A Case of Muscular Dystrophy with Dilated Cardiomyopathy: Do Not Forget Your Basics

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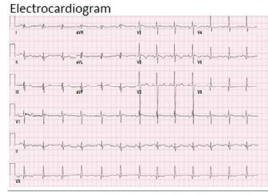
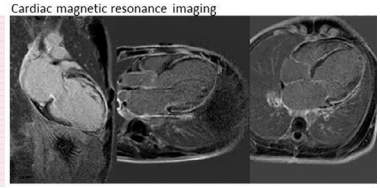
Introduction: Becker muscular dystrophy (BMD) is an X-linked recessive disorder with dystrophin mutation. Dilated Cardiomyopathy (DCM) is a leading cause of death in BMD patients. Herein, we are presenting a patient with BMD that initially sought medical attention for acute onset of systolic heart failure that highlights the importance of careful clinical assessment and appropriate work up.
Case Report: A 29-year-old male with medical history of asthma presented to the hospital with progressive dyspnea and leg swelling. He was diagnosed with DCM with an LVIDD of 6.5 cm and LV ejection fraction of 20-25% by echocardiogram. Coronary angiogram revealed no coronary artery disease. Initial blood work and electrocardiogram are below (Figure). Cardiac MRI showed severely reduced biventricular systolic function with near circumferential, sub-epicardial to mid-myocardial delayed gadolinium enhancement (Figure). Initial differential diagnosis included prior myocarditis vs. burnt out sarcoidosis. It was subsequently noted that patient began recurrently falling with muscle weakness from age 20 years with chronically elevated AST and CK. His exam was notable for atrophy of the bilateral quadriceps muscles, decreased muscular strength and bilateral calves hypertrophy. Electromyography showed evidence of chronic proximal and distal myopathy, predominantly affecting the lower extremity. Skeletal muscle biopsy showed fascicular atrophy and hypertrophy, focal endomyosial fibrosis and an increase of central nuclei without evidence of inflammation or granuloma which was most suggestive of a muscular dystrophy. Genetic testing was then completed and showed hemizygous dystrophin mutation confirming diagnosis of BMD.
Summary: BMD has a diffuse phenotype and should be considered in young patients with cardiomyopathy and chronically elevated CK and AST. A thorough clinical history, exam, and CMR can assist in directing need for skeletal muscle biopsy and subsequent genetic testing.

Figure
Electrocardiogram



Cardiac magnetic resonance imaging



	BNP pg/ml	Cr, mg/dL	AST, IU/L	ALT, IU/L	CPK IU/L	Hs-Trop ng/L
Labs on admission	1,044	0.77	98	81	847	59

BNP= B-type Natriuretic Peptide, Cr= Serum Creatinine, AST= Aspartate Transaminase, ALT= Alanine Transaminase, CPK= Creatine Phosphokinase, Hs-Trop= High-Sensitivity Troponin

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Sublingual Administration of Tacrolimus and Cases of Significant Nephrotic and Neurologic Toxicity

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Introduction: This topic warrants consideration due to the popularity of sublingual administration of tacrolimus and the possibility of deleterious effects. There are significantly different characteristics of sublingual tacrolimus compared to oral administration. In bypassing enterohepatic circulation, a greater proportion of circulating tacrolimus may be available in a 'free' form, unbound to proteins and blood cells. This is similar to a rapid intravenous delivery of calcineurin inhibitor which has noted toxicities.
Case Report: (Case 1) A 49 year old male underwent heart transplant without induction, sublingual tacrolimus was started and after two days of trough levels of 5ng/mL or below, the dose was increased to 4mg SL bid, levels increased from to 17.8 ng/mL. While creatinine increased from 1.7 mcg/dL (POD#2) to a peak of 5.1 mcg/dL on POD#6. The patient was otherwise quite stable. Tacrolimus was held temporarily then converted to oral administration. The creatinine eventually recovered to a value of 1.5mg/dL. The average tacrolimus trough level (while receiving oral dosing) during this period of creatinine recovery was 5.9 ng/ML. (Case 2) 64 year old man with ICM received a heart transplant with uneventful operative course and no induction. He received tacrolimus 1mg orally pre-operatively. Then he received 2mg Of the drug sublingually twice daily until POD # 2. On POD#3 he experienced a generalized tonic-clonic seizure that terminated with anticonvulsant therapy. The tacrolimus trough levels rose quickly after the initial tacrolimus sublingual dosing postoperatively, measured at 11.4 ng/mL on POD#2. Neurology recommended a switch to cyclosporine (goal trough level 200-250ng/mL). The patient remained on levetiracetam for 14 days, no further epileptic activity was observed. Renal function during this episode from a creatinine of 1.4 mg/dL on POD#1 to 4.2 mg/dL on POD#3.
Summary: The molecular size of tacrolimus allows for passage into the glomerulus when in the free form. This may also facilitate blood/brain barrier transport. The presented cases provide cause for consideration of possible toxicity of tacrolimus given sublingually, particularly in patients whose levels rise rapidly. Through rapid absorption, the drug may not have sufficient time to bind to proteins and cells before it reaches end organs in which toxicity occurs.

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A Closer Evaluation for Refractory Atrial Fibrillation with Heart Failure, a Case Report

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Introduction: Atrial fibrillation (AF) is highly prevalent after the age of 55 years. However, it can be associated with infiltrative cardiomyopathy.
Case Report: A 63 years old had new onset of palpitation and shortness of breath. He presented to ED with rapid AF at rate of 140 bpm treated with