

Association between Anderson-Fabry disease clinical severity and high-sensitivity troponin-T levels



Pieruzzi F^{1,2}, Salerno F¹, Di Giacomo A¹, Binaggia A¹, Torti G², Stella A^{1,2}.

1. Health Sciences Department, Milan-Bicocca University, Milano, Italy. 2. Clinical Nephrology, San Gerardo Hospital, Monza, Italy.

OBJECTIVES

While there are but a few reports of the relationship between Anderson-Fabry disease (AFD) and Troponin I (1,2), the present study is, to our knowledge, the first report of the relationship between AFD and high-sensitive Troponin-T (TnT-hs). The purpose of the study is to observe the relationship between serum levels of TnT-hs with the severity of heart and renal disease in AFD patients.

METHODS

Nineteen, clinically stable, adult patients with a genetic diagnosis of AFD underwent transthoracic echocardiography; serum TnT-hs (Roche Diagnostics), N-terminal pro-Brain Natriuretic Peptide (NTproBNP), urinary albumin / creatinin ratio (uACR), serum creatinine levels were measured, and the latter was used to estimate GFR (eGFR, CKD-EPI formula). The patients were divided in two groups on the basis of serum levels of TnT-hs, using the 99th percentile in the healthy general population as a cut-off value (14 ng/L) (3). Data are expressed as a mean \pm standard deviation for continuous variables, as an absolute value for categorical variables. Continuous variables were analyzed with Student's t-test for independent samples. The associations between continuous variables were evaluated with linear regression analysis.

RESULTS

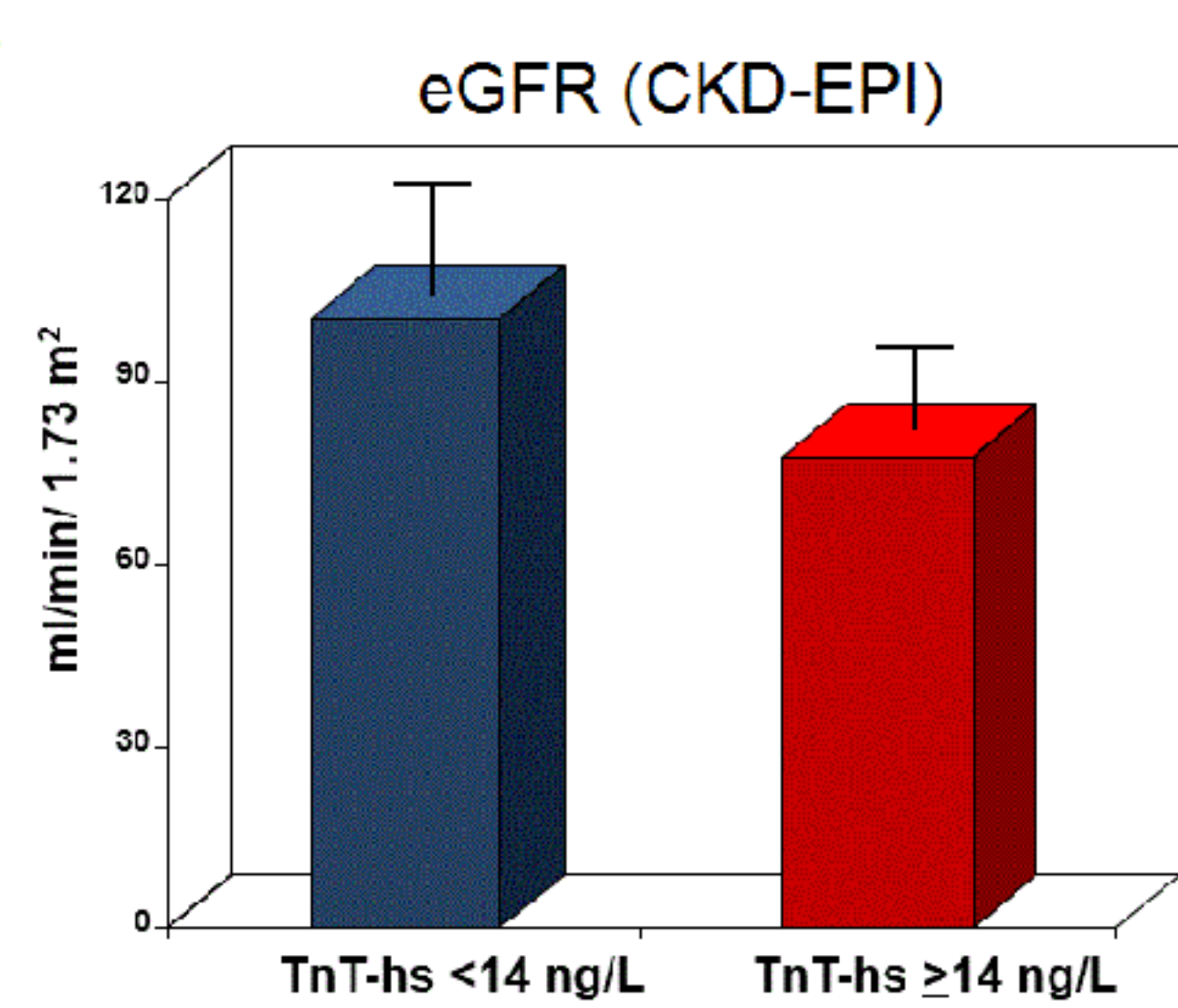


Figure 1: eGFR in patients with TnT-hs < 14 ng/L vs TnT-hs \geq 14 ng/L

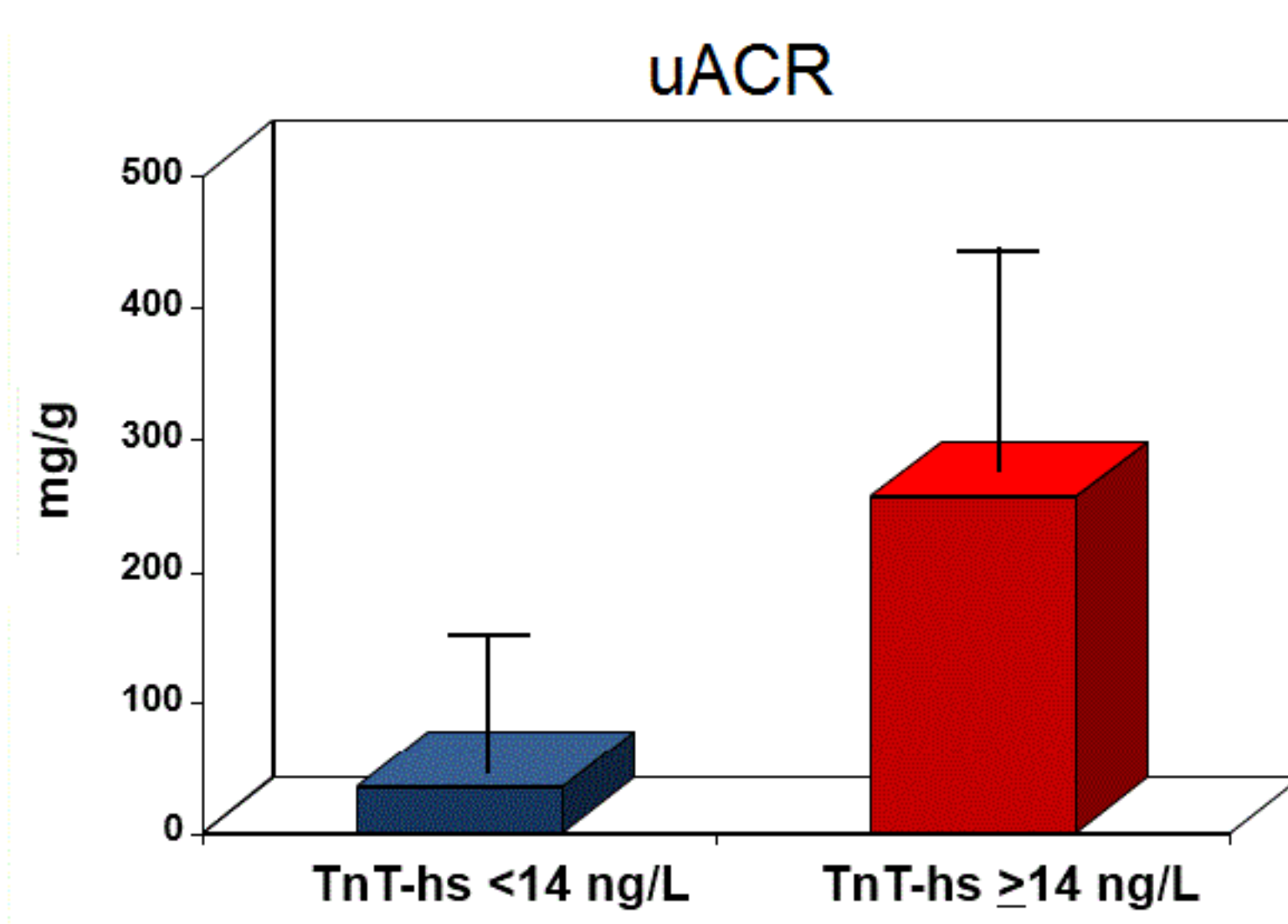


Figure 2: uACR in patients with TnT-hs < 14 ng/L vs TnT-hs \geq 14 ng/L

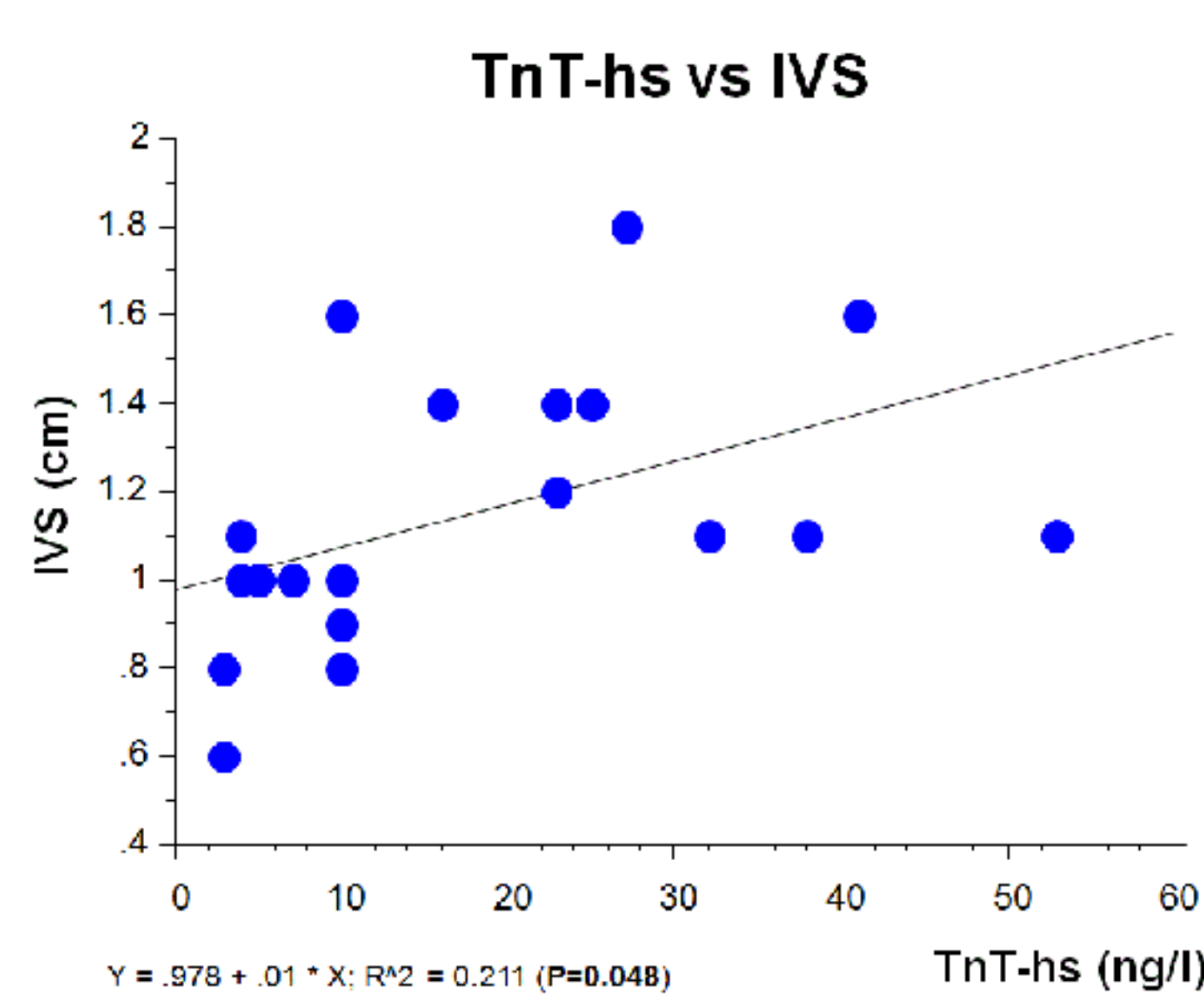


Figure 3: linear regression model for the relationship between TnT-hs and IVS thickness

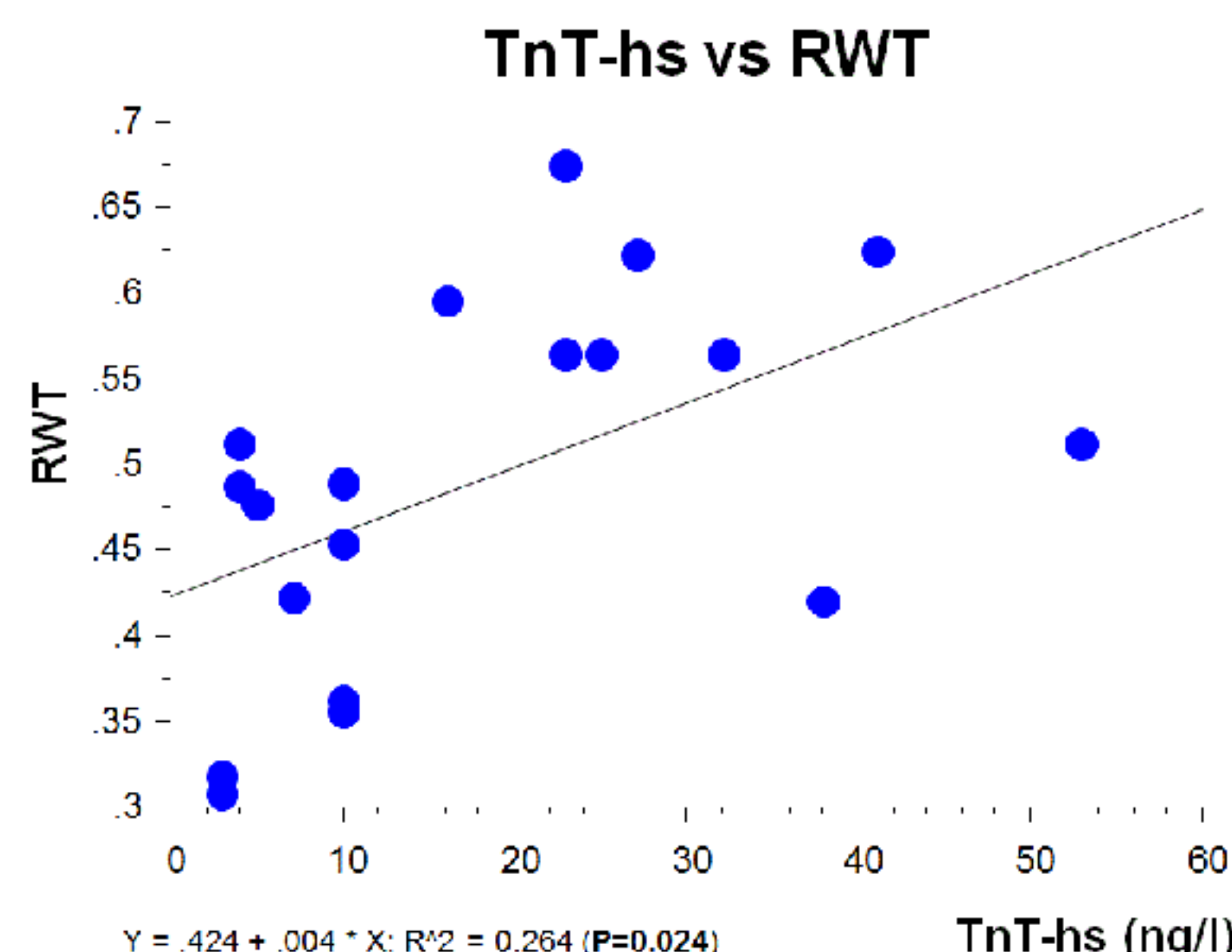


Figure 4: linear regression model for the relationship between TnT-hs and RWT

	TnT-hs < 14 ng/L (n=10)	TnT-hs \geq 14 ng/L (n=9)	P-value
Age (years)	44.4 \pm 15	53.9 \pm 8.4	0.081
Males (no)	0	5	
ERT (no)	4	7	
TnT-hs (ng/L)	6.6 \pm 3.1	52.3 \pm 67.9	
eGFR (ml/min/1.73 m ²)	100.0 \pm 15.9	77.3 \pm 27.3	0.047
uACR (mg/g)	35.5 \pm 66.2	256.9 \pm 441.8	0.041
NT-proBNP (pg/ml)	46.8 \pm 43.3	850.9 \pm 1003.0	0.029
IVS thickness (cm)	0.98 \pm 0.26	1.43 \pm 0.3	0.004
RWT	0.42 \pm 0.08	0.61 \pm 0.08	<0.001
LVMI (g/m ²)	88.8 \pm 44.8	139.2 \pm 49.7	0.049
LAD (cm)	3.5 \pm 0.8	4.1 \pm 0.7	0.11
E/Em	9.4 \pm 5.8	15.4 \pm 4.5	0.036

Table 1: patient and clinical data. ERT: enzyme replacement therapy. uACR: urinary albumin/creatinin ratio. IVS: interventricular septum. RWT: relative wall thickness. LVMI: left ventricular mass index. LAD: left atrium diameter. E/Em: E wave transmitral doppler velocity, Em tissue doppler velocity at the mitral annulus

CONCLUSIONS

High serum levels of TnT-hs are associated with an increased severity of organ involvement in patients affected by AFD. As expected, male gender is associated with more severe clinical involvement and higher serum TnT-hs levels as compared to females. Female gender, however, is characterized by wide, complex clinical variability and, in this context, higher levels of TnT-hs may be useful in defining overall clinical burden.

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