

## Isolated adrenal deficiency & Hashimoto's thyroiditis in Patient with Double outlet right ventricle

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Isolated ACTH deficiency(IAD) is a relatively rare disease, but this disease is often reported in association with autoimmune disease. We report the case of a patient who presented with aggravated biventricular function caused by Atrial fibrillation(A.Fib) associated with IAD and Hashimoto's thyroiditis(HT). A.fib with a rapid ventricular response can cause heart failure(HF) and an association with IAD would exacerbate a patient's condition. A 35-year-old men presented with palpitation, general weakness, fatigability, poor oral intake which worsened gradually over several years with weight loss. He had history of Rastelli operation, Ventricular septal defect closure, right ventricular outflow tract reconstruction with homograft due to double outlet right ventricle(DORV), pulmonary stenosis. An electrocardiogram, revealed A.fib with a ventricular rate of 126 beats/min. mild cardiomegaly was observed in chest X-ray. Echocardiography revealed decreased both Ventricular function [EF 44%, TAPSE 7.2mm, Moderate TR, RV-PA proximal Conduit 12mm, peak vel. 2.4m/s(PG 20 mmHg). In laboratory test, increased NT-pro BNP level 1282 pg/ml [0~263 pg/ml] TSH level 19.07uIU/mL[0.8~1.70 uIU/mL], normal range in free T4 1.1ng/dl[0.8~1.70 ng/dl], decreased Cortisol 0.2ug/dl [AM 6~10 6.02~18.4 ud/dl], ACTH 2.9 pg/ml [AM 8~10, 10~60 pg/ml] were reported. TPO antibody, Anti-TG antibody were positive, Anti-nuclear antibody was weakly positive of speckled pattern. Combined pituitary function test revealed IAD. There were no abnormal findings in Sella MRI, Adrenal CT and Thyroid sonography. Thus, diagnosis of IAD and Subclinical hypothyroidism due to HT was made. patient was started on Hydrocortisone 10mg/10mg, levothyroxine 25ug daily. In 10-months follow-up, patient's symptom and LV function was gradually improved and NT Pro-BNP level was decreased from 1282 pg/ml to 135.9 pg/ml. no more A.flutter was observed. We presented the case of a patient with systolic dysfunction caused by A.fib associated with IAD and HT. pathophysiology of HF and previous DORV sometimes cannot be easily explained. but treatment of hydrocortisone and levothyroxine is helpful to improve patient's LV dysfunction.

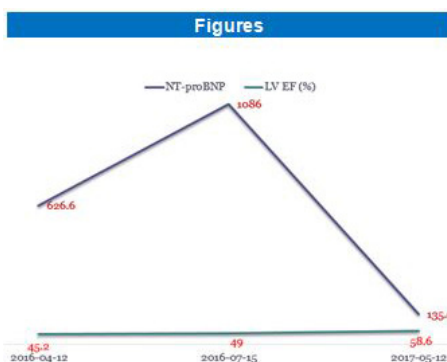


Figure 1. Clinical course and concomitant LV EF, NT-pro BNP level response to Hydrocortisone and Synthroid therapy.

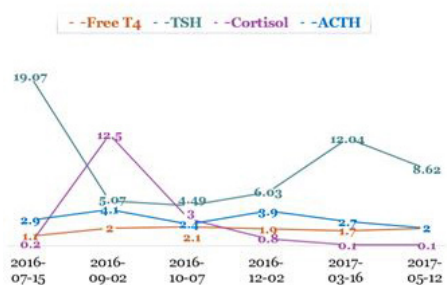


Figure 2. Clinical course and concomitant Free T4, TSH, Cortisol, ACTH levels and response to Hydrocortisone and synthroid therapy.