

Two Faces of Baroreflex Failure: Managing Hypotensive and Hypertensive Surges in Patients with Familial Dysautonomia

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INTRODUCTION

The Vagus nerve is a key part of the baroreflex network, which is a series of nerves that work to control blood pressure and heart rate. It can sense shifts in blood pressure in both directions, and reacts to prevent it from rising or falling too much. Without the vagus nerve, these reflexes to control blood pressure would not work. Familial Dysautonomia is a rare genetic disease that originated in Ashkenazi Jews in the 1500s that results in a congenital neuropathy preventing some of the cranial nerves from developing. Medical advances and research have extended the lifespan of patients with FD. 20 years ago, patients lived till the age of 5. Now, the life expectancy has increased to 40 years old and many patients are even in their 50s. Life expectancy depends on the severity of the condition and there is a wide variation of symptoms ranging from minimal ability of function to living a quality of life in school and with a job. These patients have no ability to sense and control these extreme blood pressure fluctuations due to afferent baroreflex failure. The hallmark feature of Familial Dysautonomia is hypertensive vomiting crises, which occur when the patient is fearful, angry, excited or ill. On the opposite spectrum, their blood pressure severely drops upon standing known as Orthostatic hypotension. They appear to have little to none vagal nerve function and cerebral autoregulation and therefore can't sense what is going on with their blood pressure leading to these swings from low to high based on gravity, emotions, and other factors. Pharmacological treatments aimed to fix one problem often exacerbates the other. Therefore, other patient management methods can be used to treat these patients such as positioning, taking vital signs, and having an interdisciplinary team approach including the dental team and the Dysautonomia Center.

METHODS & MATERIAL

The blood pressure variability in patients with FD may have long term implications for end organ damage as well as complications with treatment and management of the patient. This literature review includes these studies that were used to describe and evaluate the circadian pattern of blood pressure in the FD population and relationships with organ function, use of medications, and overall disability in their daily life. The assessment and management of baroreflex dysfunction are reviewed.

ORAL IMPLICATIONS

Patients with FD have several orofacial manifestations. These include dysphagia, risk of aspiration, hypersalivation, higher risk of soft tissue injury due to absence of pain, malocclusions, and postural hypotension. It is important to be mindful that patients with FD have decreased pain sensation and sensory impairment, therefore procedures such as restorations and extractions may be done without local anesthesia. Dental trauma is common due to self injury on the tongue, lips, and mucosa. Dental management includes:

- Monitor vital signs
- Keep patient in a semi supine upright position to prevent risk of aspiration
- Recline chair slowly and wait a minute before getting up due to OH
- Rubber dam isolation and suction for excessive drooling
- Hypertension management with fentanyl bolus
- Minor procedures can be done in the OR under sedation
- NSAIDS and morphine for post-op pain
- Intraoral appliances such as mouth guards can be given to reduce soft tissue injury

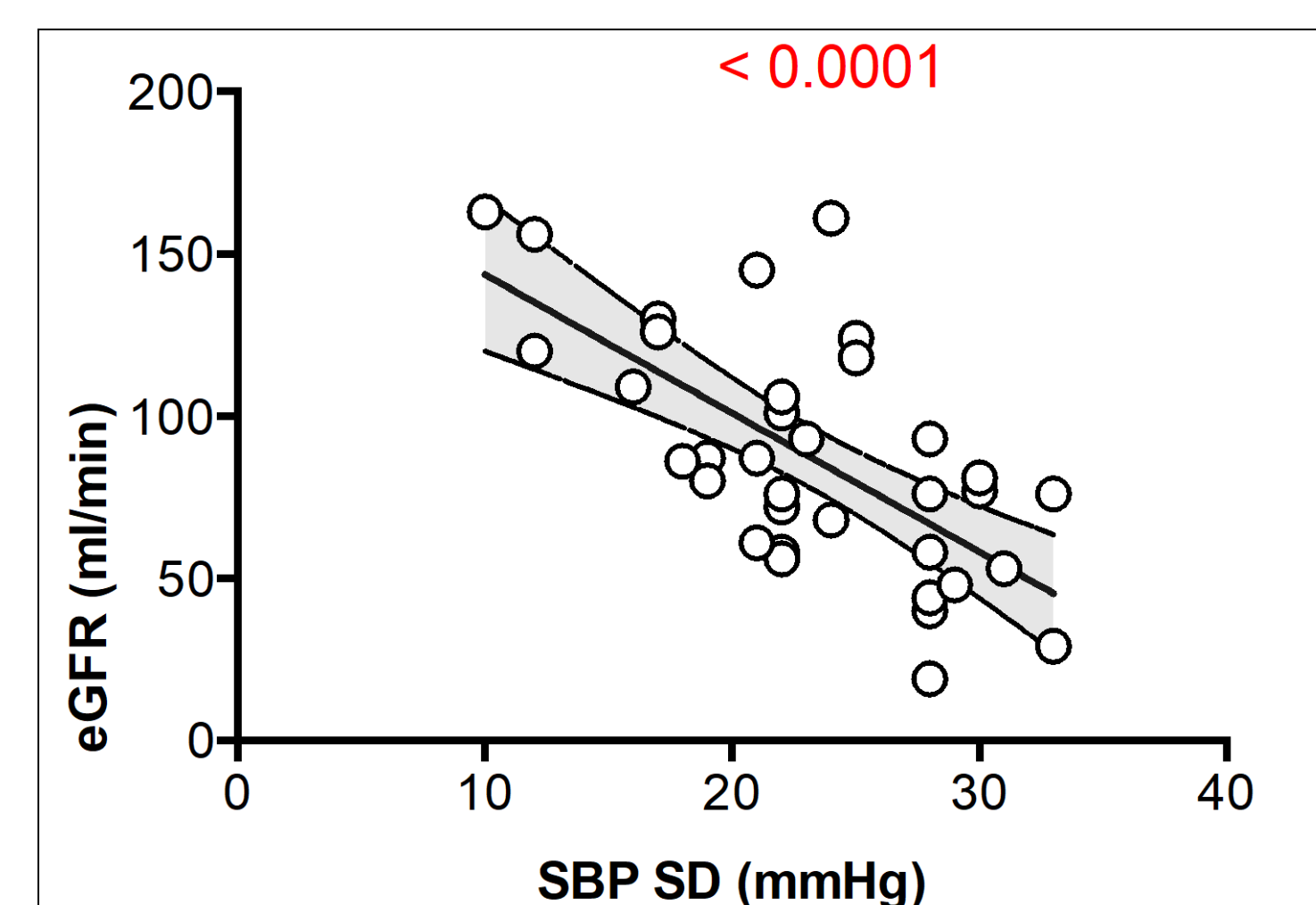
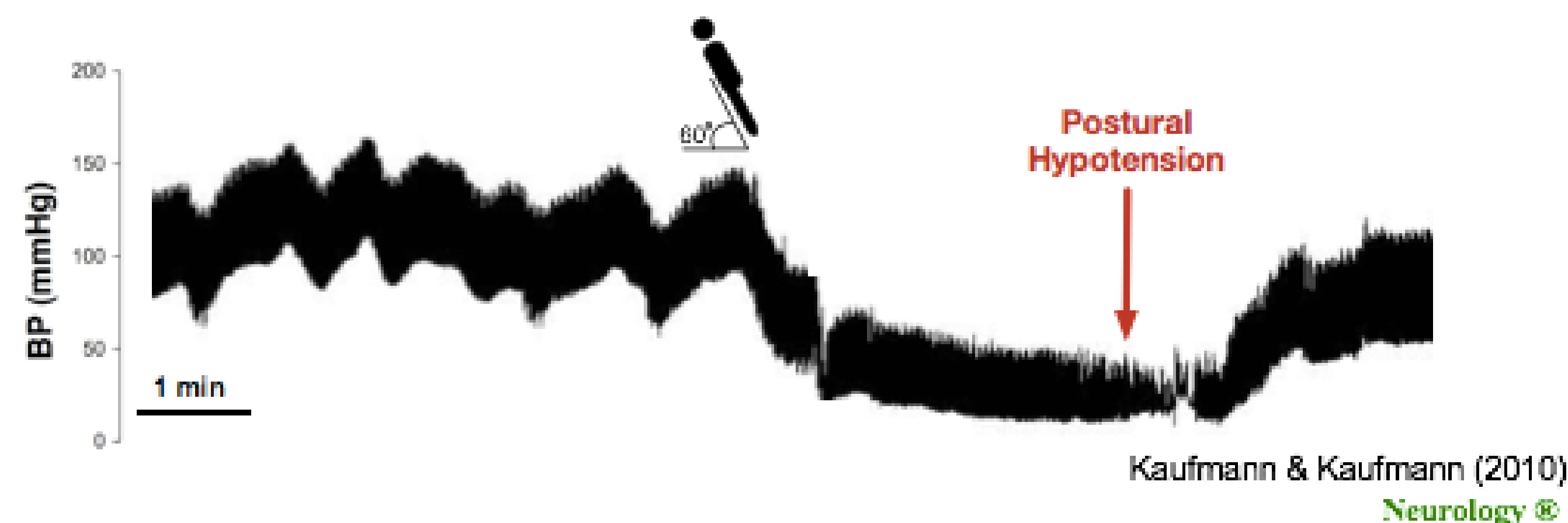
Due to the unpredictable nature of the disease and the wide spectrum and variation of symptoms these patients have, treatment planning should be individualized on a person to person basis. Frequent dental visits are recommended and both medical and dental interdisciplinary cooperation is needed for the treatment of these patients.

ABSTRACT

Familial dysautonomia is a genetic disease diagnosed at birth due to a genetic deficiency of the protein IKAP, which affects development of peripheral neurons. Patients with FD show complex abnormalities of the baroreflex receptor. This literature review examines multiple studies done to evaluate blood pressure changes in patients with FD. The autonomic responses triggered by postural changes and emotions were examined in 50 patients with FD and compared to those of normal subjects by performing tilt table testing. During upright tilt, patients with FD had severe blood pressure drops. Upon returning to supine position, blood pressure increased. Mental stress caused a marked increase in blood pressure and heart rate in patients with FD but little change in those with other types of autonomic failure and healthy controls. These findings indicate that IKAP is critical for the development of afferent baroreflex pathways and has therapeutic implications in the management of these patients.

RESULTS

In office tilt table tests as well as 24 hour ambulatory blood pressure monitor data showed that In contrast to controls, patients with FD did not have a normal response to standing. All met consensus criteria for orthostatic hypotension. Long term studies recording hypertensive surges and comparing to GFR proved that blood pressure fluctuations have negative impact on organ damage specifically the retina and the kidney.



CONCLUSION

Afferent baroreflex failure in patients with Familial Dysautonomia can cause severe supine hypertension and orthostatic hypotension, making it one of the most challenging forms of hypertension to manage. Patients lack the ability to coordinate the autonomic nervous system, so they cannot control the pressure of blood reaching the brain. After just moments of standing, blood pressure can fall to extraordinarily low values. On the other hand, emotions such as anger, excitement, anticipation of an event, fear can cause hypertensive vomiting attacks. Long-acting central sympatholytic drugs remain the main treatment. More research is needed to validate treatment recommendations and develop preventivemeasures.

The goal of treatment should not be to normalize blood pressure, but rather reduce the extreme ranges in blood pressure to improve the patient's quality of life. Mental stress is the most common trigger of hypertensive crises. These patients often have an extreme case of "white-coat" hypertension. This may respond to biofeedback techniques, benzodiazepines and cannabinoids. For orthostatic hypotension, adequate hydration as well as sodium in the diet is necessary. Research underscores the importance of measuring blood pressure in different positions. In addition to taking readings while the patient is sitting, it's also important to take measures lying flat and standing up. This range of blood pressures can help guide treatment. It's important to detect and manage supine hypertension as this has clear consequences for organ damage.