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Editorial

The Pediatric Endocrinologist's Role in the Diagnosis of Hypertension in Girls with Turner Syndrome

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Editorial

Cardiovascular disease is the major morbidity reducing lifespan in Turner Syndrome (TS) and systemic hypertension is the most readily treatable risk factor for preventing ischemic heart disease, stroke, aortic dissection and rupture [1]. Yet hypertension has few overt symptoms and can go unrecognized and untreated unless providers are attuned to the importance of appropriate screening, diagnosis and treatment in this population.

Girls and women with TS often navigate the healthcare system without much guidance. Primary care, endocrinology, cardiology, nephrology, orthopedics, audiology, and others often play important roles; however, each patient requires a different combination of specialists and focused screening to cater to their specific needs. Dedicated TS clinics consolidate some of the medical care, but many patients must individually assemble a patchwork team from the providers that are locally available.

Hypertension is common in TS with a prevalence of 25-40% in children and an even larger percentage in affected women [2-5]. A large study of girls with TS highlighted the awareness gap and under treatment of hypertension-most with elevated Blood Pressure (BP) were unaware and more than half with known hypertension had elevated BP despite treatment [6]. The causes of hypertension in TS are multifactorial, however appropriate treatment of primary and secondary causes is impossible without proper recognition [7,8].

TS vasculopathies, such as carotid thickening and arterial stiffness progressively worsen over time [9] and untreated hypertension increases the risk of aortic dilation and aortic dissection [2]. This progressive nature presents a time-sensitive opportunity for earlier intervention and makes a strong case for frequent monitoring. Yet, who is responsible for screening, diagnosing and treating hypertension in this population?

The traditional players in the screening, diagnosis and treatment of hypertension may not adequately perform this for patients with TS. Most primary care providers will follow only a few patients with TS in an entire career and may not have expertise on hypertension within the context of TS. Cardiologists, depending on a patient's cardiac anatomy, may evaluate them once every one to five years or longer. Most patients will not need to see a nephrologist unless there is an anatomical or functional renal/urologic issue. However, nearly all girls with Turner syndrome see a pediatric endocrinologist regularly for growth hormone treatment, pubertal hormone replacement, or management of autoimmune disorders. Perhaps the pediatric endocrinologist is best positioned to screen for, diagnose, and treat hypertension in girls with TS.

Several barriers interfere with effectively managing hypertension. Some are common across general pediatric care such as correct measurement technique and use of appropriate BP thresholds: for example, a BP of 112/75 may be normal for most adolescents or adults, but for a 5-year-old with a height at the 5th percentile, this is at

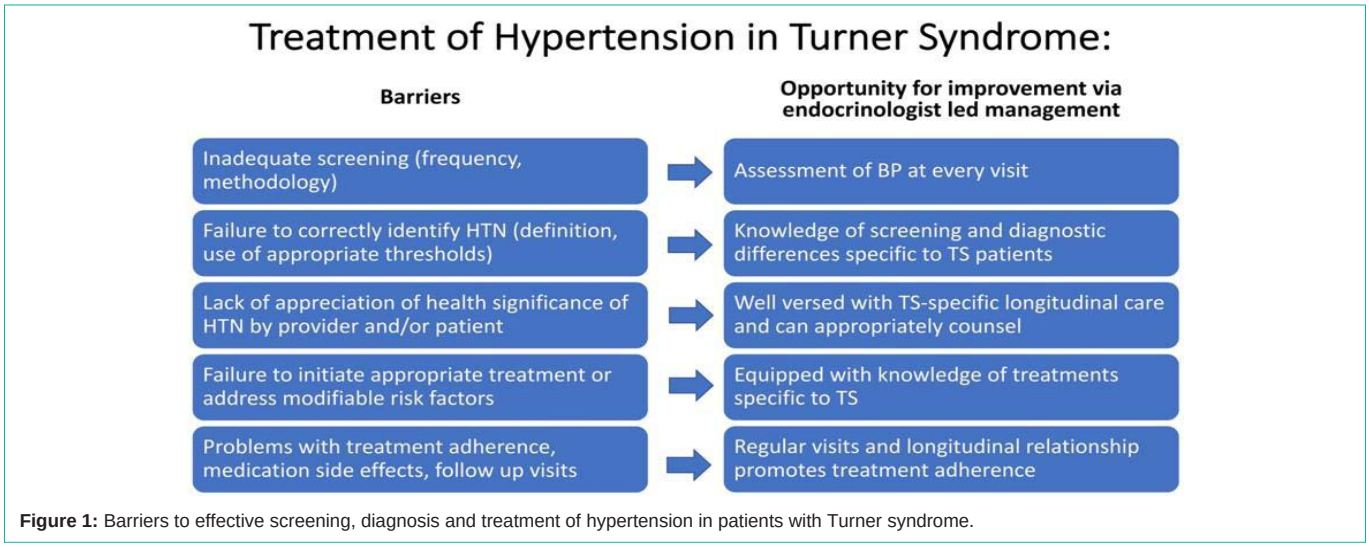

Figure 1: Barriers to effective screening, diagnosis and treatment of hypertension in patients with Turner syndrome.

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the 95th% and concerning for possible hypertension. Other challenges intrinsic to the TS population such as nocturnal hypertension may make diagnosis more challenging, or perhaps the lack of knowledge that certain anti-hypertensive drugs have the added benefit of slowing the rate of aortic dilation [10]. Finally, a specialized provider might simply view hypertension as "someone else's job" (Figure 1). Each of these potential barriers offer an opportunity for improvement in screening, diagnosis and treatment of this prevalent risk factor.

Often the pediatric endocrinologist serves as the de facto TS medical home and is uniquely equipped to address hypertension. In addition to appropriate BP screening, timely diagnosis of hypertension, discussing the significance to overall health, initiating appropriate pharmacotherapy and monitoring treatment, we must also leverage our experience and resources for weight management, which is perhaps the most modifiable risk factor for hypertension. Identifying the barriers to effective hypertension treatment empowers endocrinologists to reduce the burden of cardiovascular morbidity in our patients with TS.

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