

MANAGING RENOVASCULAR HYPERTENSION IN NEUROFIBROMATOSIS: THE ROLE OF RENAL AUTOTRANSPLANTATION

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Introduction

INTRODUCTION: Renovascular disease significantly contributes to systemic arterial hypertension in about 7% of pediatric patients with Type 1 Neurofibromatosis (NF1). Effective management often requires surgical intervention. This case report highlights the use of renal autotransplantation as a surgical approach for managing severe arterial hypertension in a child with NF1 and provides insights from a systematic literature review on the subject.

Methods

A systematic literature review was conducted in October 2024 following PRISMA guidelines, focusing on surgical management strategies for renovascular hypertension in pediatric patients with NF1. Searches were performed in databases such as PubMed, EMBASE, and Cochrane CENTRAL, targeting studies that explored surgical interventions, specifically renal autotransplantation. Additionally, a case study of a two-year-old girl with NF1 and uncontrolled hypertension was analyzed.

Results

A two-year-old girl with NF1 presented to the Emergency Department with critically high blood pressure. Despite multiple antihypertensive medications, her hypertension remained uncontrolled. A renal ultrasound with Doppler raised suspicions of left renal artery stenosis, leading to further evaluations, including renal scintigraphy, urinary catecholamine measurements, and renal arteriography. Normal urinary catecholamine levels ruled out pheochromocytoma, while arteriography confirmed severe left renal artery stenosis.

Given her persistent hypertension despite treatment with three antihypertensive drugs, surgical intervention was indicated.

The patient underwent resection of the stenotic segment and reimplantation of the left renal artery into the intrarenal aorta. The procedure was uneventful, and follow-up renal Doppler ultrasound showed no signs of stenosis, indicating hemodynamically stable arteries.

Consequently, antihypertensive medications were gradually reduced, leading to normalized blood pressure levels. The systematic review identified several case reports and small series demonstrating the efficacy of renal autotransplantation in managing complex renovascular conditions in NF1 patients, with favorable long-term outcomes for blood pressure control and renal function preservation.

Conclusions

This study illustrates that renal autotransplantation is a viable surgical option for pediatric patients with NF1 experiencing severe renovascular hypertension. The systematic review supports the effectiveness of various surgical strategies for this condition, particularly when traditional methods like percutaneous transluminal angioplasty may not be suitable. Early identification and surgical correction of renal artery stenosis can effectively preserve renal function and prevent hypertension-related complications, underscoring the importance of awareness among pediatric healthcare providers.

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